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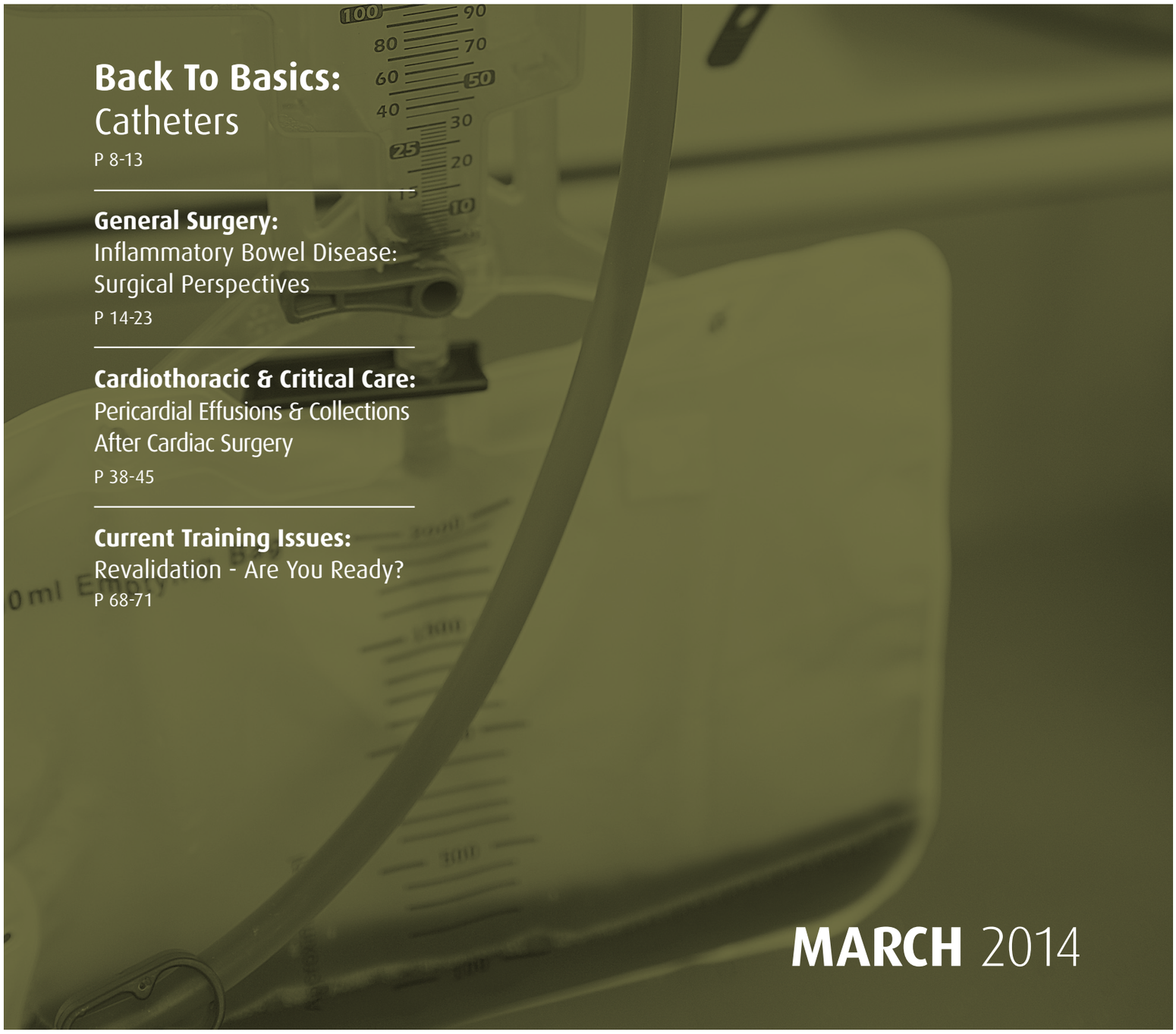
Volume 4, Issue 2



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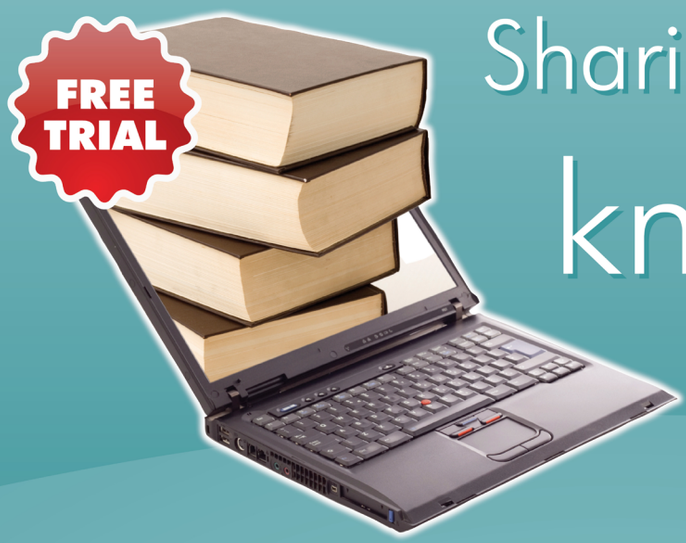

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Dear Prospective Author,

Thank you for considering the submission of an article to 'Core Surgery'. Our journal aims to educate and inform junior surgical trainees about relevant 'core' subject topics. Each issue will cover a topic from selected subspecialty fields; General Surgery, Orthopaedics and Trauma, Plastic Surgery, Ear Nose and Throat Surgery, Neurosurgery, Urology, Paediatric Surgery and Intensive Care Medicine. Articles will be required to be broad enough to help with preparation for the intercollegiate MRCS examination but also focus on key hints and tips on becoming a higher surgical trainee.

A list of core topics in each subspecialty has therefore been agreed by the editors based on a selection of key topics in the MRCS curriculum. Authors are advised to agree a topic with the editors before writing an article. We strongly recommend that all articles have a senior author of registrar level or above.

Types of Article

Manuscripts are considered under the following sections:

1) Case based discussions
2) Practical procedures
3) Review articles
4) Research papers
5) Back to basics
6) Careers focus: Current training issues, course reviews, audit, charitable experiences, career pathway

Submission of Manuscript and Covering letter

Submissions will only be accepted via email and must be accompanied by a covering letter bearing the corresponding author's signature. Please submit your article to **coresurgery@123doc.com.** The covering letter must contain an acknowledgement that all authors contributed significantly and are in agreement with the content of the manuscript. In addition any financial or other conflict of interest must be declared. All submissions must be accompanied by a electronic copies of the transfer of copyright and conflict of interest disclosure forms (see below).

Papers are accepted for publication in the journal on the understanding that the content has not been published or is being considered for publication elsewhere; this must be stated in the covering letter. If authors submit manuscripts relating to original research, the corresponding author must state in the covering letter that the protocol for the research project has been approved by a suitably constituted ethics committee and that it conforms to the provisions of the Declaration of Helsinki (as revised in Brazil 2013), available at **http://www.wma.net/en/30publications/10policies/b3/**

All investigations involving human subjects must include a statement that the subject gave informed consent and patient anonymity should be preserved. This statement must be included in the covering letter and duplicated at the end of the main manuscript.

Conflict of interest

All authors must complete an individual conflict of interest declaration form which should be downloaded from **http://www.icmje.org/coi_disclosure.pdf** and submitted electronically to **coresurgery@123doc.com.** Instructions for completion can be found at the ICMJE website as above: **http://www.ICMJE.org/.**

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Submissions should follow the style of the Vancouver agreement detailed in the International Committee of Medical Journal Editors' revised 'Uniform Requirements for Manuscripts Submitted to Biomedical Journals: Writing and Editing for Biomedical Publication', as found at **http://www.ICMJE.org/**

References

All articles must be referenced appropriately. The Vancouver system of referencing should be used; details can be found at **https://workspace.imperial.ac.uk/library/Public/Vancouver_referencing.pdf.** References should be cited using bracketed numerals in the order in which they appear e.g. (1). The list of references should reflect this order and names of journals should be abbreviated in the style used in Index Medicus **ftp://nlmpubs.nlm.nih.gov/online/journals/ljiweb.pdf.**

Format of Articles

Guidelines for the format of respective article types are as follows. All articles must contain an abstract of 150-250 words, and must include up to five keywords for indexing purposes. A title page must be included containing the title of the article and full details for all authors including forename, initials, surname, specialty, grade, institute and email address. A contact postal mailing address should also be supplied for the corresponding author who should be separately identified.

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Case based discussions

Should be about 1000-1500 words long and should focus on clinical assessment, differential diagnosis or treatment. The basic structure should be as follows:

- **Abstract: The salient points of the case and discussion.**
- **Case history: Including the initial presentation, clinical setting and problem, investigation and treatment.**
- **Discussion: Covering the critical aspects of the management and the treatment options.**

Practical Procedures

Should be about 1000-1500 words long. Although not essential it is highly advantageous if pictures and diagrams are supplied to illustrate the most salient points. Articles should be set out as follows:

- **Abstract (Essential) –A summary of the article structure and salient features.**
- **History and pathology**
- **Indications and contraindications**
- **Gaining informed consent/ explaining procedure to patient**
- **Equipment required**
- **Draping / sterile field preparation**
- **Patient positioning and relevant anaesthetic points**
- **Documentation of procedure / recording of complications and management of such**

Review articles

The topic should be relevant to core surgical trainees, and a maximum of 2500 words long. The review should include an abstract, and a clinical vignette of a case relevant to the topic. The aim of including a clinical case is to provide a focus for discussion, and to ensure that the review is relevant and useful to our readership.

Research papers

Although the publication of research articles is not a core aim of the journal, Core Surgery welcomes research submissions if thought to be of interest to the readership. Articles should be written using the following headings (title page, abstract, introduction, methods, results, discussion, references). They should be a maximum of 2500 words of text including abstract, 30 references, 3 illustrations or figures. The abstract should be a maximum of 250 words and use the following headings (introduction, methods, results, conclusion). The title page should contain the title of the paper, the full names of the authors, the addresses of the institutions at which the research was carried out and the full postal address, email address and telephone number of the corresponding author.

Back to basics

These are articles covering basic principles and practice of surgery and should include general topics pertinent for the core surgical trainee. A topic or subject should be agreed with an editor prior to submission; please email **coresurgery@123doc.com** for further details. Articles should be a maximum of 1500 words long and must include an abstract.

Careers focus

Articles in this section may include course reviews, audits, description of charitable experiences, discussion of current training issues, and 'career pathway' articles providing an overview of training with hints and tips for aiding progression. Topics should be agreed with an editor prior to submission; please email **coresurgery@123doc.com** for further details. Course reviews should describe a course which is either mandatory or desirable for core trainees and junior higher surgical trainees. Audits should preferably be those where the cycle is complete, or have led to guideline development.Each article must contain an abstract.

MCQs / EMQs (All Articles)

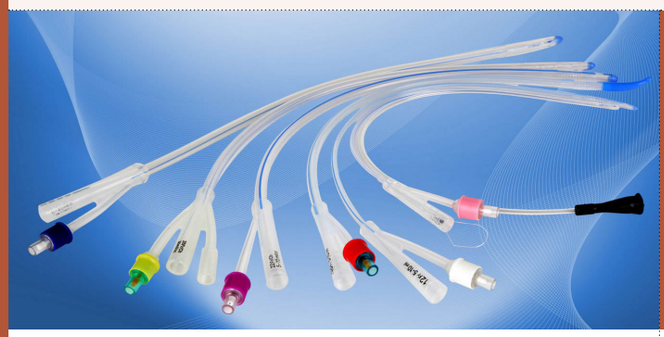
Please note that all articles should be submitted with five multiple choice questions (MCQs) or extended matching questions (EMQs) attached, in the style of the Member of the Royal College of Surgeons (MRCS) 'Part A' examination. These questions should have answers and brief teaching notes/discussion included in a separate paragraph following the questions. Examples of the requirements for question style can be found here: **http://www.intercollegiatemrcs.org.uk/new/pdf/part_a_sample_mcqs.pdf**

Summary

Articles considered for publication will be sent for review by our panel of consultants and junior surgical trainees. We wish you every success with your submission. Please contact the editorial team with any questions.

CATHETERS

CJ Ridd, A Gulamhusein



Abstract

Urinary catheterisation is a common procedure in many hospital and community patients with more than 30 million catheterisations being carried out worldwide each year (1). It forms one of the required core competencies for medical students and foundation doctors in the UK. Knowledge of catheterisation is part of the MRCS syllabus (2) and the skill has been examined in the MRCS Part B OSCE. This article discusses types of catheter in common use, indications for insertion, catheterisation technique and related complications. It also aims to provide practical solutions for common catheter related difficulties a surgical trainee is likely to encounter.

Keywords: catheter, urethral, suprapubic, Foley, technique

Introduction

The use of urinary catheters, indications and risks can be dated from the 5th century BC (3). The well recognised self-retaining Foley catheter was designed by Dr. Frederick Eugene Basil Foley, an American urologist who first described its use in 1929 for patients post cystoscopic prostatectomy.

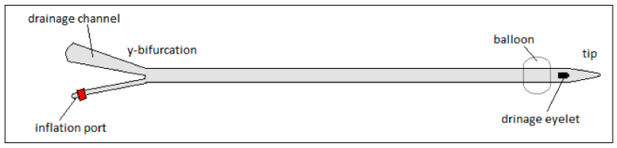


Figure 1: Foley Catheter.

Types of urinary catheter

The design and composition of urinary catheters has evolved greatly. The original Foley catheters were made of latex rubber, however, although cheap these have largely become obsolete due to the high incidence of infection and hypersensitivity reactions. Current catheters in regular use are more commonly all silicone or latex coated in silicone, PTFE (polytetrafluoroethylene) or hydrogel. With the exception of PTFE coated, these are all classed as long-term catheters and can remain in-situ for up to 12 weeks. The PTFE coating tends to damage over time and as a consequence these medium-term catheters need changing at 28 days (4).

Catheters are also available impregnated with silver alloy and/or antibiotics. These catheters aim to reduce catheter associated urinary tract infections (CA-UTI), however current evidence suggests that they do not significantly reduce morbidity and are not cost effective (5, 6).

Choice of catheter depends on its purpose. One-way catheters only have one channel for drainage, with no balloon as they are not intended to remain in the bladder for a long period of time. They are commonly used for intermittent catheterisation and instillation of intravesical drugs. Two-way catheters are most common and have one channel for urine and another for inflation of the balloon. Three-way catheters have a third channel to enable continuous bladder irrigation, generally used following urological surgery or to clear blood clots and debris within the bladder.

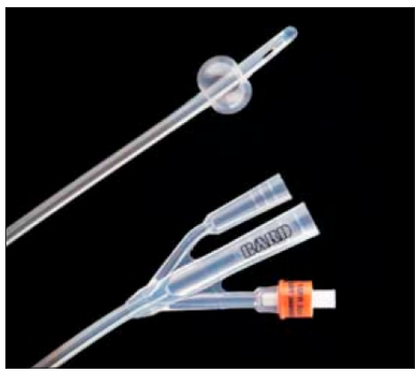


Figure 2: 3-way catheter (courtesy of BARD Medical).

Catheter size is measured in Charrière (Ch) also known as French Gauge (Fr) and indicates the external diameter. 1mm = 3Ch and therefore a 12Fr catheter has an external diameter of 4mm and an approximate circumference of 12mm. In general, smaller catheters are more comfortable for the patient and it is recommended to use 12-14Ch for females and 14-16Ch for males.

CATHETERS

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Urethral Catheterisation

Indications for urethral catheterisation:

Emergency

- *Acute urinary retention.*
- *High pressure chronic urinary retention.*
- *Need for accurate measurements of urinary output (post-operatively or in acutely unwell patients).*
- *For bladder irrigation or lavage.*

Elective

- *Patients undergoing urological surgery.*
- *Peri-operative use in selected surgical procedures e.g. prolonged duration.*
- *Maintain bladder drainage for patients with neurological voiding dysfunction.*
- *To ensure continence and preserve skin integrity.*

Contraindications to urethral catheterisation

- *Acute prostatitis.*
- *Suspected urethral trauma.*

• Explain procedure and gain patient consent.
• Check patient allergies.
• Prepare trolley with all necessary equipment.
• Position the patient appropriately.
• Women: supine lithotomy position with legs apart & knees flexed.
• Men: supine, legs slightly apart.
• Wash hands, put on sterile gloves and an apron.
• Clean the introitus/glans with an appropriate cleaning solution.
• Drape with sterile drapes.
• Identify the meatus and introduce 10-15mls of lubricating gel. • Ensuring ample lubrication is key, particularly in difficult male catheterisations.
• Make sure the end of the catheter is lubricated and prepare to insert the catheter into the external urethral meatus.
• Women: the urethra is a short membranous canal (4cm), advance caudally until urine is seen in the catheter.
• Men: Apply traction on the penis and hold it vertically to straighten the urethra. Difficulty may be encountered as it traverses the external sphincter and then again as it negotiates past the U-shaped bulbar urethra. At this point, lower the penis to a horizontal position and apply steady, gentle pressure. Ask the patient to strain gently as if to void urine which may be helpful.
• Once urine flows freely from the catheter advance it a bit further to ensure it is within the bladder.
• Inflate the balloon with the recommended volume depending on the catheter. Pain on inflation may suggest the balloon is within the urethra and should therefore be deflated and the position re-evaluated.
• Attach the catheter bag and ensure the catheter moves freely in the urethra.
• Document the residual volume and the appearance of the urine drained.
• In non circumcised men replace the foreskin to avoid a paraphimosis.

Technique Box 1 : Urethral Catheterisation.

Difficult Catheterisation

In the female patient, and especially in post-menopausal women, difficulty catheterising is usually related to difficulty in defining the anatomy.

To increase your chance of successful catheterisation get assistance, put the bed at a comfortable height, ensure the area is well lit and that the position of the patient is optimal. Put the patient in the trendelenburg position or place a pad underneath the buttocks to lift the pelvis. Take time to identify the anatomy; the meatus is located in the introitus, below the clitoris and above the vaginal orifice. The meatus can be hidden in a skin fold. Gently wipe with a swab or probe possible sites with the catheter tip to locate the meatus. Otherwise feel for the meatus by running a finger along the anterior surface of the lower vaginal wall in the midline (7).

In the male patient, difficult catheterisation can be due to an inability to access the meatus, meatal stenosis, urethral stricture, prostatic enlargement, a high bladder neck or the presence of a false passage.

Figure 3: Male Urethral catheterisation Dummy (courtesy of Creative Commons Attribution – Share Alike License).

In a patient with phimosis you can try lifting the foreskin forward to visualise the meatus. If this is not possible you can attempt to gently insert the catheter blindly. Remember that the normal position of the meatus is slightly inferior to the horizontal midline in the centre of the glans. If the phimosis is too tight the patient may require a dorsal slit. (7)

When the penis is buried, visualise the penis by having an assistant press firmly around the base. Hewes et al. described inserting a flexible cystoscope into the tract leading to the penis, allowing passage of a guidewire under direct vision, which a catheter was then fed over (8). Penile oedema can be reduced with compression for up to 20 minutes prior to further catheterisation attempts. Dilating sounds can be used in patients with meatal stenoses; however this is rarely tolerated under local anaesthesia.

CATHETERS

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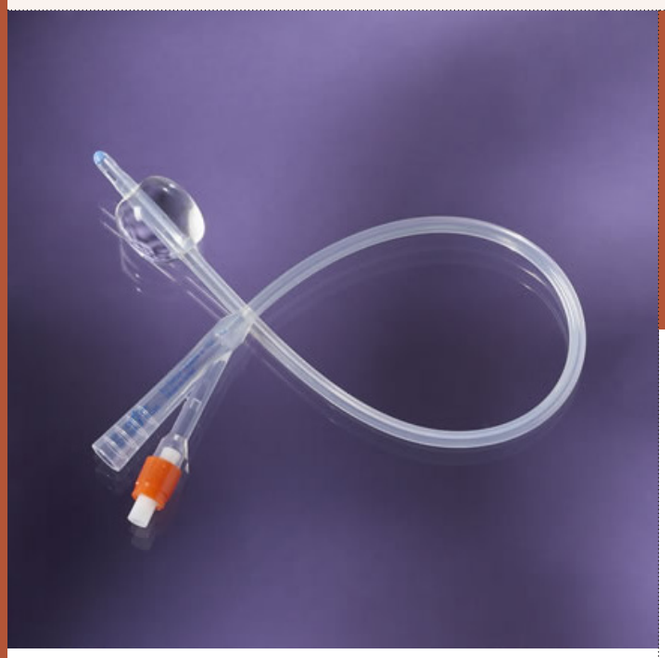

If you are meeting resistance as the catheter negotiates the bulbar and prostatic urethra, then surgical trainees with experience and training can utilise a curved tip catheter, commonly a Coudé or Tiemann tipped catheter. In a survey of American urology residents swapping to a Coudé tip enabled catheterisation of approximately 40-50% of patients where a standard catheter could not be passed (9). When using a catheter with a curved tip, keep the tip of the catheter at 12 o'clock to facilitate passage around the prostate gland.

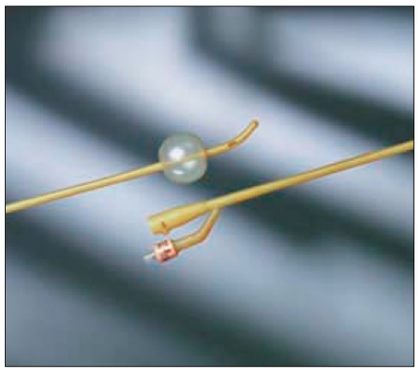

Figure 4: Tiemann Tip Catheter (Courtesy of BARD Medical).

If you suspect a urethral stricture try using a smaller catheter. Due to the increased rigidity it is often easier to pass an IC than the equivalent size Foley in patients with strictures. Be wary of forming a false passage.

If the above measures fail the patient will need a urology referral. At this point, often the patient will undergo a flexible cystoscopy with a view to catheter insertion over a guide wire, depending on the underlying problem.

Removing a catheter

To remove a catheter insert a syringe into the valve and the syringe should automatically fill as the balloon deflates. This process can be aided by gentle pressure on the syringe but be mindful that strong pressure can collapse the inflation channel. Ensure the balloon is fully deflated before removing the catheter with gentle backward traction.

When it is not possible to deflate the balloon, the options below can be tried.

- **Try dislodging the obstruction by inflating the balloon further with air or water.**
- **Leave a syringe firmly in the valve and come back after an hour. Gravity will aid the deflation process.**
- **Squeeze the visible tubing to dislodge crystal formation within the channel.**
- **Cut the balloon tube just proximal to the inflation valve, water may simply drain through the channel if the valve was faulty.**
- **If these methods fail seek urology input with a view to percutaneous puncturing of the balloon generally under ultrasound guidance (USS).**

Technique Box 2: Tricks for deflating a catheter balloon.

After catheter removal it is important to ensure the patient is adequately voiding. A trial without catheter (TWOC) involves measuring consecutive post void residual scans (generally 3 readings) to ensure satisfactory bladder emptying.

CATHETERS

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Intermittent Catheterisation

Intermittent catherisation (IC) is an option in patients who have high post void residuals or as an alternative to a long-term indwelling catheter (IDC). If the patient is able and willing, it negates the need for catheter bags and reduces the incidence of UTIs when compared to an IDC. Whereas insertion of an IDC should be under aseptic conditions, IC only requires a clean technique.

Figure 5: Lofric intermittent catheter (courtesy of Wellspect Healthcare).

Complications

False passages can be created during attempts to catheterise. If you believe a patient has a false passage which is preventing insertion of a catheter and you have some experience it is appropriate to have one further attempt with a curved tip catheter. As false passages are usually created posteriorly or laterally the curved tip may help to bypass it. Patients who have had traumatic catheterisations should be covered with antibiotic prophylaxis.

Urinary tract infections are the most common catheter associated complication and they cause significant morbidity and mortality (5, 6). It is important to distinguish between CA-UTI and asymptomatic catheter associated bacteriuria. All catheterised patients will have positive urine dipstick tests and are likely to grow organisms when their urine is sent for culture. Asymptomatic patients with bacteriuria should not be treated with antibiotics for a UTI as this only serves to develop resistance. However in the presence of sepsis in a patient with a long-term IDC you should consider the resistance profile of organisms on previous urine cultures when making an appropriate antibiotic choice.

If a catheter becomes blocked you can flush it to relieve the obstruction and if this fails replace the catheter. If the blockage is due to haematuria and clot retention you will need to insert a larger catheter to allow for regular bladder washouts or a 3-way catheter to allow bladder irrigation. Bladder irrigation can be difficult, and risky, on a ward not experienced in its use. Unless obviously traumatic (e.g. post insertion), the underlying cause for haematuria should be investigated with upper tract imaging and a flexible cystoscopy once the urine is clear.

Bladder spasm related to a urethral catheter is generally as a consequence of the balloon resting on the trigone. Ensure the catheter is draining and if so a low dose antimuscarinic is often effective. Urine bypassing the catheter can be due to blockage, infection or bladder spasm.

Long-term urethral catheters cause pressure necrosis of the urethra; in women this causes the urethra to become patulous, this means that the catheter bypasses and frequent expulsion of the catheter even with the balloon inflated can occur. In men, the catheter can erode the glans penis ventrally, resulting in an acquired hypospadias. It is therefore recommended that suprapubic catheters (SPC) be used in patients requiring long-term IDC.

Suprapubic Catheterisation

Insertion of an SPC is indicated in urinary retention when passage of a urethral catheter is not possible and is preferred in patients requiring long-term IDC. SPC insertion is absolutely contraindicated in the absence of an easily palpable or USS localised distended bladder. Relative contraindications include known or suspected carcinoma of the bladder, uncorrected coagulopathy, previous lower abdominal surgery, ascites, abdominal wall infection and prosthetic devices in the lower abdomen e.g. a hernia mesh.

SPC insertion is commonly performed using the Seldinger technique but can also be performed as an open procedure, especially where there is considered to be a risk of causing bowel perforation. If there has been previous lower abdominal surgery or the bladder is not palpable USS guidance must be used to ensure that the catheter is being inserted below the peritoneal reflection (10). In addition cystoscopy can be used to fill the bladder and this is the preferred method in patients undergoing elective SPC insertion.

Although SPC insertion is carried out routinely, the associated morbidity (bowel perforation 0.15 -2.7%) and mortality (0.54 - 1.8%) is significant. The British Association of Urologists (BAUS) published guidelines in 2010 which set out recommendations to improve the safety of SPC insertion (10).

Ideally the first SPC change should happen 8-12 weeks after insertion allowing the tract time to mature. Changing an SPC is straightforward but some resistance can be felt as the balloon cuff passes through the abdominal wall. If an SPC is removed unintentionally, assuming the tract is mature (>2 weeks post initial insertion), immediate endeavours should be made to recatheterise the tract as it will close quickly. Immediate urology referral is recommended if this is not possible.

CATHETERS

CJ Ridd, A Gulamhusein

Catheters
Back to Basics

MCQs

1. Which of the following is an absolute contraindication to blind SPC insertion:

a) Haematuria

b) Bladder not palpable

c) Ascites

d) Previous lower abdominal surgery

e) Spinal injury

2. Which of the following is not an indication for urethral catheterisation:

a) high pressure chronic retention

b) measuring urine output in an acutely unwell patient

c) to allow bladder irrigation/lavage

d) managing intractable incontinence

e) Heavy haematuria and clot retention following a road traffic accident

3. A 27 year old male spinal injuries patient with paraplegia and chronic urinary retention is best managed with:

a) long-term urethral catheterisation

b) transurethral resection of the prostate

c) intermittent self catheterisation

d) long-term suprapubic catheterisation

e) treatment with an alphablocker and 5 alpha reductase inhibitor

4. A 70 year old female patient presents with haematuria and a history of passing clots. On admission she is in obvious distress with a palpable bladder on examination. What is your initial management:

a) insertion of a suprapubic catheter

b) insertion of a 3-way catheter

c) flexible cystoscopy to rule out a bladder carcinoma

d) insertion of a large bore 2-way catheter

e) CT abdomen and pelvis

5. Which of the following is not recommended when a catheter balloon fails to deflate for catheter removal:

a) instil further water or air to try and displace an obstruction

b) squeeze the tube to dislodge crystal formation within it

c) leave the syringe on the valve and return after 40 minutes

d) inflate the balloon further to burst it

e) cut the balloon tube just proximal to the valve

CATHETERS

CJ Ridd, A Gulamhusein

Answers

1 - B

In this scenario the only safe way to proceed would be using USS guidance or insertion under cystoscopic guidance, whereby the bladder can be adequately filled.

2 - E

On the contrary, this is a contraindication due to the possibility of a urethral injury. A urethrogram should be performed, although if in retention an open cystotomy would be indicated.

3 - C

If physically able this is the gold standard treatment and is associated with lower infection risks when compared to IDC. His retention is not secondary to prostatic enlargement.

4 - B

This patient is likely in clot retention and therefore requires drainage of her bladder. A 3-way catheter is less likely to block and allows continous bladder irrigation. Haematuria investigations should be instigated following this initial management.

5 - D

There is a risk of remnants of the balloon remaining within the bladder which may sometimes require open surgery to remove.

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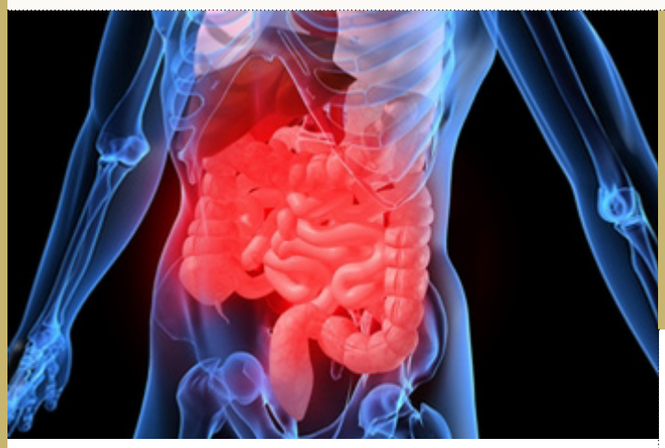
INFLAMMATORY BOWEL DISEASE: SURGICAL PERSPECTIVES

I Hamzah, I Sheldrake



Abstract

Inflammatory bowel disease is a complex spectrum of gastrointestinal disease that forms a significant bulk of work for both gastroenterologists and general surgeons. The approach in managing this disorder requires careful consideration of the constantly changing management paradigms. Having an exceptional pathophysiological understanding of this disease is paramount, but great emphasis is now towards a more multidisciplinary approach with surgeons, physicians and the patients themselves involved in the decision making. In this article, we will discuss about the clinical presentation and its various diagnostic modalities, followed by the medical and surgical management of inflammatory bowel disease.

Keywords: *Crohn's disease; Inflammatory bowel disease; Ulcerative colitis.*

Clinical vignette

A 21-year-old man presents to the emergency department with peri-anal pain and discharge. He reports a 2-month history of intermittent bloody diarrhoea with associated loss of weight of about one stone over a month. His abdomen is soft, but slightly tender on palpation in the right lower quadrant. On examination, he is apyrexial with normal vital signs. The examination of the rectum is difficult to perform due to pain, but an area of erythematous swelling is visible close to the anal margin, discharging watery pus from its apex. Several anal tags are also present.

Introduction

Ulcerative colitis (UC) and Crohn's disease are the main subtypes of idiopathic inflammatory bowel disease (IBD). They are chronic inflammatory diseases defined by clinical, histological, endoscopic and radiological features (1) which typically follow a relapsing-remitting pattern. There are about 240,000 IBD sufferers in the UK of which 145,000 have UC (2) with the incidence of Crohn's disease significantly increased from 1950s until 1980s (3). Since then, incidence has markedly reduced (2). Both diseases can manifest themselves at any age, but peak incidence is in the 20s and 30s.

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General Surgery

Aetiology and pathology

UC is characterised by diffuse mucosal inflammation affecting the rectum which extends proximally to varying degrees but is limited to the colon. Superficial mucosal ulceration is common and the abundance of neutrophils within the lamina propria and crypts form micro-abscesses (1).

Crohn's disease results in macrophage aggregation which forms non-caseating granulomas. The terminal ileum is most commonly affected, but any part of the gastrointestinal tract can be involved. In contrast to UC, Crohn's inflammation is transmural but often patchy and segmental (1). The aetiology of Crohn's is largely unknown. The inheritable component is stronger than in UC (4) and the pattern of increased worldwide incidence suggests a contribution of environmental factors. Smoking has also been shown to predispose to Crohn's, but protects against UC through unknown mechanisms (5).

The Montreal classification system is used to accurately categorise the various IBD phenotypes and patterns of inflammation (6). 5% of patients with inflammatory changes in the colon have clinical, radiological, endoscopic and pathological features of both diseases. This is termed 'IBD, type unclassified (IBDU)'. 'Indeterminate colitis' refers to post-colectomy patients who still do not fit either classification after full pathological examination (6).

Clinical features including extra-intestinal features

Approximately 50% of UC sufferers will have at least one relapse per year, with about 80% being mild/moderate and 20% severe (2). A quarter of patients with UC will suffer one or more episodes of acute severe colitis in their lifetime with a third requiring colectomy (2). Management of acute severe colitis has improved over the years but it is still life-threatening, with a mortality of up to 2%. When the disease is active, patients experience urgency to defecate, colicky abdominal pain, tenesmus with bloody diarrhoea as the cardinal symptom.

Crohn's disease sufferers experience abdominal pain, diarrhoea and weight loss, with systemic symptoms such as malaise and fever. Complications such as strictures, fistulae, abscesses and obstruction are also more common with fistulae occuring in approximately one quarter of Crohn's sufferers (7). Unlike for UC, surgery is not curative and morbidity is greater with only 75% of patients fully capable of work 1 year after diagnosis compared to 90% of UC patients.

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Extra-intestinal symptoms are present in approximately 21-40% IBD sufferers (8). Most studies have shown these to be more common in Crohn's than UC, with colonic Crohn's disease patients having the highest prevalence (9). Anaemia is the most common extra-intestinal manifestation. Dermatological complications include erythema nodosum, which is most common in females with Crohn's, pyoderma gangrenosum, and apthous stomatitis (10).

Musculoskeletal symptoms include asymmetrical seronegative arthropathy which can affect up to 30% of IBD sufferers as well as sacroilitis and osteoporosis. Ocular complications include anterior uveitis, scleritis and episcleritis which respond well to treatment of the underlying bowel condition. Hepatobiliary complications including steatohepatitis and primary sclerosing cholangitis (PSC), which is strongly associated with UC (5% UC patients have PSC, and 70% of patients with PSC have UC (11)). Nephrolitiasis is associated with Crohn's disease by causing increased oxaluria secondary to disruption of terminal ileum function.

Diagnosis and investigation

A comprehensive look by the British Society of Gastroenterologist have led to the production of a guideline that provide clinicians with an educated and evidence-based support in the management of inflammatory bowel disease (12). A full history, with particular attention to recent travel and infections, medications, smoking and family history should be taken. Information on stool consistency, frequency, urgency, evidence of bleeding, abdominal pain, malaise, fever, weight loss and any extra-intestinal manifestations should be gathered. Examination should include measurement of all physiological parameters alongside abdominal and perineal examination. Blood tests should include FBC, UE, LFT, CRP, ESR, ferritin, vitamin B12 and folate.

Faecal calprotectin can accurately detect colonic inflammation. Stool samples must be sent for culture and C. difficile toxin testing as IBD sufferers become infected more readily. At least 4 samples are required to detect 90% cases (13). Plain abdominal radiography must be performed to exclude dilatation and identify proximal constipation.

Colonoscopy with numerous biopsies is first line in the diagnosis of colitis. In acute severe colitis, sigmoidoscopy can be more readily performed. Rectal biopsies must be taken even if no macroscopic changes are apparent. Biopsies attempt to define the type of IBD, reveal coexisting complications such as infection, and identify any evidence of dysplasia, grading it appropriately.

Ultrasound cannot be used in isolation, but is radiation-free, and sensitive for detecting disease in the terminal ileum. Operator subjectivity limits its clinical value.

CT has a role in rapidly assessing patients for complications such as obstruction or abscesses and is currently the 'Gold Standard'. For imaging of the small bowel, MRI enterography is the recommended technique but current availability limits its use. However, availability is improving, and this test is replacing barium small bowel meal and CT as the preferred method of imaging the small bowel.

The IBD Service Standards state there should be no more than a 4 week wait for outpatient radiological or endoscopic investigations. Inpatients with severe exacerbations should wait no longer than 24 hours and biopsy results should be available within 5 days.

The risk of developing colorectal cancer for people with inflammatory bowel disease is estimated as 2% after 10 years, 8% after 20 years and 18% after 30 years of disease for ulcerative colitis, whilst for Crohn's disease the risk of developing colorectal cancer is considered to be similar to that for people with ulcerative colitis with the same extent of colonic involvement. The National Institute for Health and Care Excellence (NICE) have issued a guideline on the colonoscopic surveillance of patients with inflammatory bowel disease, stratifying the frequency based on the risks (14);

- **Low risk:**

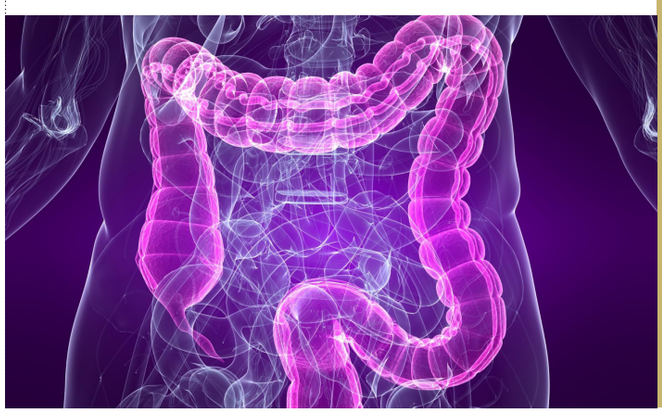
- extensive but quiescent ulcerative colitis or,
- extensive but quiescent Crohn's colitis or,
- left-sided ulcerative colitis (but not proctitis alone) or Crohn's colitis of a similar extent.
- offer colonoscopy at 5 years from last complete colonoscopy.

- **Intermediate risk:**

- extensive ulcerative or Crohn's colitis with mild active inflammation that has been confirmed endoscopically or histologically or,
- post-inflammatory polyps or,
- family history of colorectal cancer in a first-degree relative aged 50 years or over.
- offer colonoscopy at 3 years from last complete colonoscopy.

- **High risk:**

- extensive ulcerative or Crohn's colitis with moderate or severe active inflammation that has been confirmed endoscopically or histologically or,
- primary sclerosing cholangitis (including after liver transplant) or,
- colonic stricture in the past 5 years or,
- any grade of dysplasia in the past 5 years or,
- family history of colorectal cancer in a first-degree relative aged under 50 years.
- offer colonoscopy at 1 year from last complete colonoscopy.



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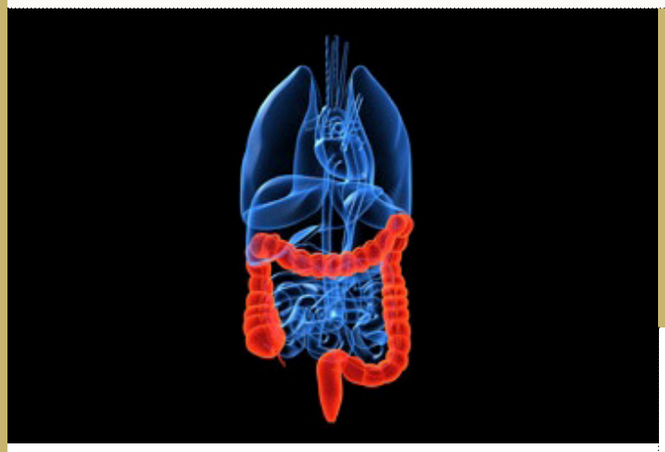

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Medical management

Truelove and Witts criteria

The Truelove and Witts criteria is used widely in clinical practice to classify the severity of UC and can be used to predict response to medical management (15). The severity may guide the various methods and medications available to manage IBD before surgical intervention becomes necessary. They fall into 4 main categories; lifestyle, maintenance, immunosuppression and rescue therapies.

	MILD	MODERATE	SEVERE
Bowel movements/day	<4	4-6	>6
Blood in stools	Small amounts	Mild-severe	Visible blood
Pyrexia (>37.8°C)	No	No	Yes
HR >90bpm	No	No	Yes
Anaemia	No	No	Yes
ESR (mm/hr)	<30	<30	>30

Table 1: Truelove and Witts criteria for inflammatory bowel disease.

Lifestyle - Nutrition, smoking, NSAIDs

Nutritional support and advice plays an important role in the management of IBD. A secondary lactose intolerance can occur in active colitis. A dairy-free diet may relieve flatulence, bloating and colickly pain. In patient with ileal Crohn's, micronutrients such as calcium, fat-soluble vitamins, zinc, iron and B12 should be monitored annually in view of their impaired absorption (16). If the absorption of the gut is reduced dramatically, short bowel syndrome may occur. In such cases, TPN may be required. About 20% of patients on TPN have Crohn's as their underlying condition (17). The use of prebiotics is unproven, but probiotics have been shown to be as effective as mesalazine in maintaining remission in UC (18). There is, however, no evidence to support their use in Crohn's disease.

Smoking cessation in Crohn's has been shown to reduce the risk of relapse by two thirds and reduce the risk of needed a surgical resection (19). The use of NSAIDs in not established, with many conflicting studies. Low dose aspirin is currently thought to be safe in the short term.

Maintenance

5-ASA

5-Aminosalicylic acid (5-ASA) has a key role in the maintenance of remission of UC. Rectal 5-ASA is also superior to rectal steroids in inducing remission in mild-moderate UC (20). In addition, it has been shown to reduce the risk of colorectal cancer by 75% (21). The use of 5-ASA in patients with Crohn's disease has shown only modest effects and is therefore not first line therapy. Nephrotoxicity is the most serious side effect and as such caution should be taken in patients with pre-existing renal disease and creatinine levels should be monitored annually.

Antibiotics

Antibiotics are important in the management of secondary complications of IBD. Ciprofloxacin, however, has been shown to be effective in the treatment of perianal Crohn's and pouchitis (22). There is no evidence to support the use of antibiotics in uncomplicated UC.

Immunosupression

Steroids

Corticosteroids are used in moderate-severe relapses of both diseases, but they have no role in maintenance therapy of either. In UC, oral prednisolone induced remission of 77% patient with mild-moderate UC compared to 48% treated with 5-ASA. A combination of oral and rectal steroids have been shown to be superior to either used individually (23). In Crohn's, a 92% remission rate has been reported at 7 weeks with no dose tapering. However, most patients relapsed quickly, with only 44% remaining in remission at 1 year, 36% required further steroids, and 20% developed steroid resistant disease (24). Escalation of therapy should be considered in patients who have severe relapses, require 2 or more courses of corticosteroid in 1 year, or relapse as the steroid dose is reduced below 15mg.

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Thiopurines

Thiopurines such as azathioprine (AZA) or mercaptopurine (MP) are widely used as adjuncts and corticosteroid-sparing therapies by inducing T-cell apoptosis. AZA is superior to 5-ASA at inducing remission in steroid dependent UC and should therefore be first line in such cases (25). In Crohn's disease, thiopurines are effective at both induction and maintenance of remission. One study quoted the odds ratio for maintaining remission with AZA was 2.43 (26). However, adverse events are common, occurring in up to 20%, including allergic reactions, hepatotoxicity and bone marrow suppression which can lead to profound leucopaenia. Some studies have also demonstrated a fourfold increased risk of developing lymphoma (27).

Methotrexate

Methotrexate is currently a second-line immunosuppressive agent which should only be used in patients resistant or intolerant of AZA or MP (28).

Rescue therapy

Ciclosporin

Ciclosporin prevents clonal expansion of T-cells and is a rapidly effective rescue therapy for patients with steroid refractory UC who would otherwise have to undergo surgery. Its use is limited to only 3-6 months due to its nephrotoxic and neurotoxic adverse effects. Opportunistic infections such as pneumocystis carinii pneumonia are well-documented complications. Ciclosporin is of no therapeutic value in Crohn's disease and has no role for patients who have relapses whilst taking AZA.

Anti-TNF

Two biological agents are currently licensed for use in IBD in the UK. Infliximab (IFX) and Adalimumab (ADA) are both monoclonal antibodies against TNF-Ð. Both can be used in Crohn's disease which has not responded to immunosuppression and studies have shown an 81% remission rate at 4 weeks compared to 17% from those who took a placebo (29). IFX in also effective in fistulating Crohn's causing resolution of fistulae in 36% versus 19% on placebo (29).

Only IFX is effective in UC, with symptom relief achieved in 69% versus 37% taking a placebo (30). There is an increased risk of non-Hodgkin's lymphoma for patients on anti-TNF therapy, but the overall risk is small (30). There are also reports of exacerbations of demyelinating disorders. IFX and ADA are contraindicated in congestive cardiac failure.

Surgery in inflammatory bowel disease

Ulcerative colitis

Whilst medical treatment serves as first line, the risk of surgical intervention in ulcerative colitis in the form of a colectomy is approximately 30%, ranging from 5% to 50 % (31, 32). Nevertheless, the option of surgery should be taken within a multidisciplinary setting with the close involvement of colorectal surgeons and gastroenterologists to ensure optimum management with close input of the patient. Nevertheless, the option of surgery essentially falls under these broad indications; severe fulminating disease irresponsive to medical intervention, chronic illness and high risk dysplastic changes.

In severe diseases, evidence of free perforation, generalised peritonitis and massive colonic haemorrhage indicates the need for emergency surgery. Surgery is also indicated in the deterioration of acute colitis (increasing toxicity or colonic dilatation) at any time after initiation of adequate medical management or if there has not been a clear improvement within 24–72 hours of admission. Development of toxic megacolon is usually an indication for early surgery. About half the patients with acute fulminating colitis respond to medical therapy, thereby avoiding emergency surgery. The majority of these patients develop repeated episodes of toxic dilatation or incapacitating chronic symptoms, ultimately requiring surgery.

In cases of protracted diseases with associated chronic complications may be amenable to elective surgery as these subgroups respond poorly to medical treatment and is troubled by recurrent acute exacerbations. The threshold for surgery by gastroenterologists and patients is variable but with the advent of sphincter-preserving restorative proctocolectomy, surgery is now better accepted. In cases where patients have severe extra-intestinal manifestations, surgery may become a considered option.

Dysplasia serves as the most sensitive marker of premalignancy, and the presence of dysplasia from a villous or polypoidal lesion or from a stricture may be an indication for prophylactic surgical management. In situations where there are synchronous lesions that indicate severe dysplasia at two separate sites in the colon will ultimately require surgery.

Laparoscopic approach in bowel resection for UC is a safe and viable option. A systematic review has shown that the overall complication rate was significantly lower for patients in the laparoscopic group compared to those who had it through an open approach (33).

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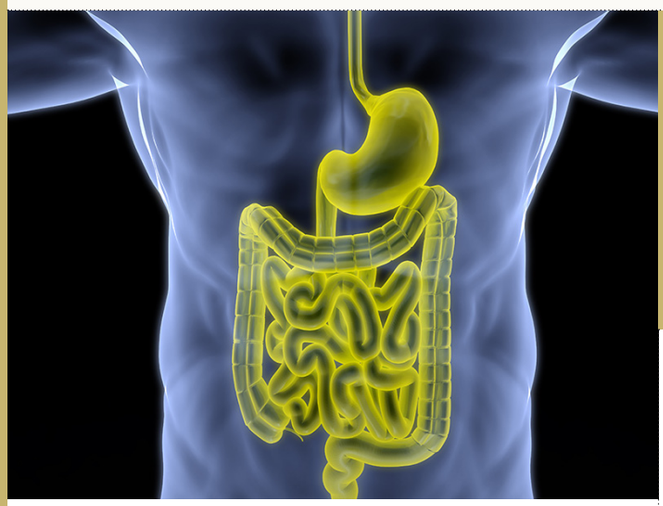

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Subtotal colectomy

Subtotal / total colectomy, end ileostomy and preservation of a long rectal stump serves as a viable option in the surgical management of ulcerative colitis within an emergency setting in patients with acute severe colitis not responding to medical treatment; as evidence suggests that prolonged medical treatment in an acute attack that leads to a delay in surgery increases morbidity (34-36). In such settings, patients are often severely unwell with poor nutritional status (seen as low albumin) and are usually on steroids; therefore primary anastomosis is not recommended at this stage. The rectal stump can either be over-sewn and left in the peritoneal cavity or brought out to the abdominal wall as a mucous fistula. It is advised that the anastomosis to restore the bowel continuity should be performed after period of convalescence with evidence of concurrent active colitis or infection resolved.

In the subtotal colectomy / total colectomy, it is the combination of right, transverse amd left hemicolectomy in which the caecum, ascending, transverse, descending and sigmoid colon are mobilised gently by incising the paracolic gutters. It is paramount to ensure that the bilateral ureters and gonadal vessels are identified and preserved during the mobilisation. Once adequate mobilisation is achieved, lymphovascular pedicle ligation can be performed starting from ileocolic, right colic, middle colic and inferior mesenteric vessels. The superior rectal artery may be preserved if a long rectal stump is intended to be left for future procedures. Ligation of the mesentery can be close to the bowel wall but as this requires multiple ligation it is preferable to choose a more proximal site. If there is high index of suspicion for dysplasia or occult malignancies (especially in long standing colitis), a more radical lymphadenectomy with vascular ligations at the mesenteric root is advisable.

A rectal stump that is left long can either be over-sewn and left intraperitoneally, or brought out as a mucous fistula. The latter may be advisable in severe colitic cases as a severely diseased rectum may perforate. An end ileostomy can then be fashioned at the ileal portion.

Restorative proctocolectomy and ileo-anal pouch procedures (IAPP)

Proctocolectomy involves the removal of the entire colon and rectum with the creation of an ileal pouch and anastomosing it to the anal canal using a hand-sewn of stapled technique. Prior to procedure, the extent of colonic disease and anal sphincter function is assessed by manometry and colonic biopsies.

In the operation itself, the entire colon is mobilised (as total colectomy) with corresponding lymphovascular supply ligated accordingly. If there are suspicions of cancer, total mesorectal excision (TME) is performed. However, if condition is strictly benign, posterior TME can be opted with meticulous identification and preservation of the pre-sacral nerves that is paramount. Once rectum is completely mobilised, transanal digital evaluation with the tip of the finger is done to mark the level of transection. It is best to transect the rectum at the top of the anal columns for a stapled anastomosis to preserve anal sensory epithelium leaving a 1 – 2 cm anal transitional zone.

There are different types of ileo-anal pouches; J, S and W with volumes ranging between 200 to 400 ml with the J-pouch being the most common. However, the key to a successful pouch is the creation of a tension-free anastomosis and it is crucial that the small bowel mesentery is mobilised adequately so that the ileal pouch will reach the levator floor with no tension.

The relative technical ease of J-pouch creation makes it the technique of choice for most surgeons; it is constructed from the terminal 30 to 40 cm of small intestine which is then folded into two 15 or 20 cm segments. An enterotomy of 1.5 cm is made longitudinally at the apex of the pouch with a subsequent side-to-side anastomosis of the two segments of the ileum using a linear stapler (2 cartridges of ILA 100) via the enterotomy at the pouch apex. A blind loop of the J-pouch is closed using a linear stapler and can be reinforced by continuous sutures. A purse string suture is applied at the apical enterotomy using a prolene suture.

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The S-pouch is constructed using 3 limbs of 12 to 15 cm of terminal small bowel with a 2 cm exit conduit with the ileum segments approximated using continuous seromuscular sutures. An S-shaped enterotomy is performed and a continuous full thickness running suture are applied to the two posterior anastomotic lines. The anterior wall is then closed with continuous seromuscular sutures which is then reinforced using interrupted sutures.

The creation of the anastomosis can either be performed via a stapled or hand-sewn technique, with the former being the most popular due to the ease and speed. A meta-analysis comparing hand-sewn and stapled anastomosis (37) showed no significant difference in post-operative complications, sexual dysfunction and quality of life between both groups. Nevertheless, the hand-sewn group had greater number of cases of liquid stool incontinence with significantly lower resting and squeezing pressure in the anorectal manometric measurements compared to the stapled group.

When comparing between the type of pouches used (J, S and W), a meta-analysis (38) perfomed showed that there were no significant differences in post-operative complications between the three group including strictures, pouchitis, leaks, pouch failure or small bowel obstruction. Events of incontinence and seepages were also comparable between the three groups. Nevertheless, J-pouches were associated with higher bowel frequency and use of anti-diarrhoeal drugs compared to the other two types. The S and W-pouches were also associated with greater difficulty in pouch evacuation which required intubation.

A practice associated with restorative proctocolectomy and IAPP is the creation of a diverting loop ileostomy by bringing the ileum proximal to the pouch to the right lower abdominal wall. The closure of the loop ileostomy can be done approximately 3 months down the line after a water-contrast pouchogram or pouchoscopy to confirm the integrity of the pouch. A meta-analysis comparing two groups of those with a loop ileostomy and those without showed that the latter group had significantly higher leak rate compared to the former. Nevertheless, those with a stoma had higher occurrence of anastomotic stricture and failure of the pouch. Events of perianal sepsis, pouchitis and pouch-related sepsis were comparable between the two groups.

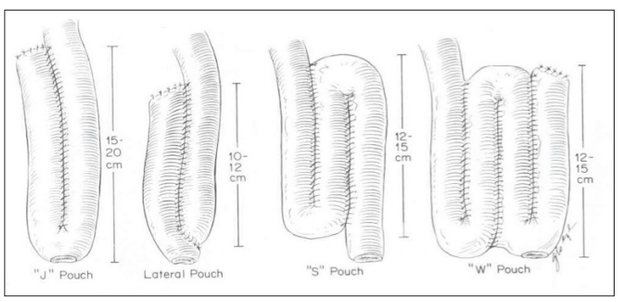


Diagram 1: Types of pouches for ileo-anal pouch procedures.

Crohn's Disease

In contrast to ulcerative colitis, Crohn's disease can affect any part of the alimentary tract from mouth to anus, and any form of surgery will not eradicate any chances of recurrences. 75% of Crohn's disease sufferer may require some form of surgical intervention within 10 years of diagnosis . The decision to undertake surgery in managing Crohn's disease is similar to ulcerative colitis. Like ulcerative colitis, surgical intervention in dictated by the severity of the disease with great emphasis on a multidisciplinary approach. However, surgery in Crohn's disease does not offer a curative outlook in patients. Resections are primarily limited to the affected macroscopic level of the bowel involvement and, coupled with the usage of stricturoplasty, have played a crucial part in the surgical management of Crohn's disease in recent years. As UC, laparoscopic approach in the surgical management of Crohn's disease is an option, although many studies performed to assess its benefits have yield conflicting outcomes in addition to most of them being non-randomized. Nevertheless, a recent meta-analysis have shown some evidence of decreased perioperative complications such as wound infection, post-operative ileus and respiratory complications as well as reduced incidence of incisional hernia in the laparoscopic group (39).

Surgical operations

Ileocolic resection

As mentioned earlier, it is best to be conservative in the surgical approach in Crohn's patients requiring surgical intervention, limiting resection to diseased areas. Ileocolic resection remains the most common surgical procedure in such patients where terminal ileal disease is the most prevalent and requiring the resection of ileum, caecum and portions (or the entire) of the right colon, which can be termed as right hemicolectomy. In this operation, the caecum and ascending colon is dissected from posterior abdominal wall up to the hepatic flexure. It is crucial that the right ureter, gonadal vessels and duodenum are identified and preserved. The entire hepatic flexure is mobilized and ileal bands are divided so that the whole of the right colon can be lifted from the abdomen. Vessels to be ligated involve the ileocolic artery and vein which is clamped and divided in the middle of the mesentery. The right colic vessels and the right branch of the middle colic vessels are also divided.

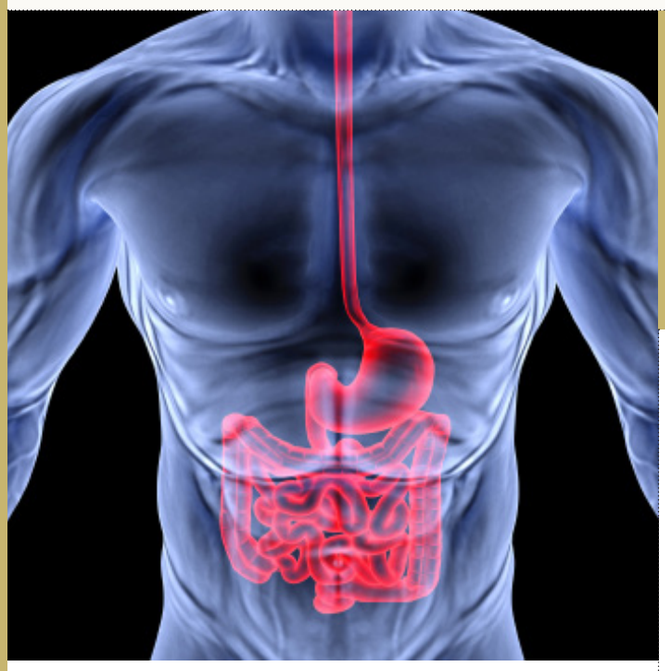
In the resection of the ileum, it is best to remove 5-10 cm of a macroscopically normal ileum proximal to the lesion. Primary anastomosis between the ileum and colon can be performed using a hand-sewn or stapled technique.

Stricturoplasty of Crohn's disease of the small bowel

Although segmental resection of diseased small bowel is an option for Crohn's disease affecting the region, stricturoplasty should be considered in cases where strictures are present but remains asymptomatic, as recurrent segmental resections may lead to complications associated with short-bowel syndrome. Stricturoplasties may also be safely undertaken concomitantly with resection depending on the presentation and the extent of the disease. Table 2 shows a summary of the indications and contraindications of this technique in small bowel disease.

INFLAMMATORY BOWEL DISEASE: SURGICAL PERSPECTIVES

I Hamzah, I Sheldrake



Indications	Contraindications
• Multiple strictures • Patients with small bowel syndrome • Previous significant small bowel resection • Strictures without associated phlegmon or fistula	• Preoperative malnutrition • Perforated bowel • Multiple strictures over short length of bowel • Bleeding from planned stricturoplasty site • Stricture short distance from area of resection

Table 2: Indications and contraindications for stricturoplasty.

There are three approaches in stricturoplasties; Heineke-Mikulicz, Finney and the side-to-side isoperistaltic stricturoplasty, but the most important factor that drives the method opted is the length of the stricture. Table 3 shows the type of stricturoplasties performed in relation to the length.

Stricturoplasty	Stricture Length
Heineke-Mikulicz	Short strictures (< 10 cm)
Finney	10 – 20 cm
Side-to-side isoperistaltic	Long strictures or multiple

Table 3: Types of stricturoplasty in relation to stricture length.

For Heineke-Mikulicz stricturoplasty, the method is confined to short segmental strictures and it involves a longitudinal enterotomy made over the antimesenteric border of the small bowel which is extended 1 to 2 cm to the other side of normal bowel. The enterotomy is then closed transversely with interrupted seromuscular sutures that are absorbable which can be done either in one or two layers ensuring that tension is avoided.

For intermediate length strictures, the Finney stricturoplasty is opted. A stay suture is placed at the midpoint of the stricture followed by an enterotomy made through the stricture extending 1 to 2 cm into the normal bowel. The stricture is then folded onto itself forming a U-shape with the posterior edges sutured in a continuous fashion with an absorbable suture. The anterior edges are then closed using interrupted sutures.

The side-to-side isoperistaltic stricturoplasty was first described by Michelassi (40) for longer strictures that extend up to 25 cm. This method involves lifting up the whole length of the strictured bowel and the mesentery for the region is divided at the midpoint. At the midpoint of the stricture, the diseased bowel is divided between atraumatic bowel clamps place to avoid spillage. The two cut ends of the bowels are brought side-to-side fashion, and then the two loops of bowel are approximated with a single layer of interrupted non-absorbable sutures.

A longitudinal enterotomy is formed on both loops for the length of the stricture with the ends of the loops spatulated to avoid blind ends or pockets. The inner layer is then approximated with a running, full-thickness absorbable suture and continued anteriorly with a running Connell stitch. The anterior layer is followed by a layer of interrupted non-absorbable seromuscular sutures.

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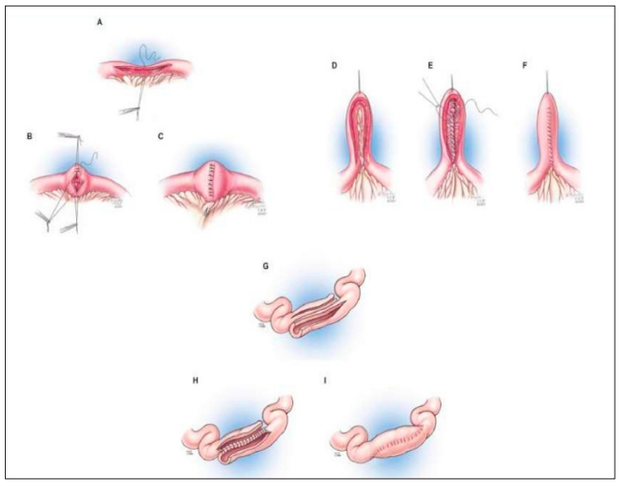

Diagram 2: Stricturoplasties (A to C – Heineke-Mikulicz, D to F – Finney, G to I – Side-to-side).

Perianal Crohn's disease: fistula-in-ano

The common approach to its management is the laying-open. A grooved probe is passed along the tract, onto which the incision is made through the anoderm, skin, fat and distal sphincter muscles to the track itself. To avoid bridging over during healing, the track is marsupialized by trimming the edges. The track is left open to granulate. This method is viable for most intersphincteric fistulae in which the distal internal sphincter is divided but the proximal internal sphincter and the entire external sphincter is preserved.

Seton placement is also another treatment option. In loose seton placement, a suture material, such as a soft braided nylon is passed through the track after which the two ends are knotted together outside the anus to form a loose encirclement. This ensures a continuous drainage of any discharge whilst preventing the premature closure of the external opening which serves as the underlying culprit of recurrent anal sepsis. Simple removal of a loose seton after a long period of good drainage should result in definitive healing in some patients.

Some patients may suffer the severe effects of perianal Crohn's and may present with recurrent perianal sepsis, incontinence and stenosis. In protracted cases that poorly respond to initial medical treatment, surgery may be an indication; faecal diversion through an ileostomy to provide bowel rest may be considered in the initial stage, failing which a proctectomy is considered to ensure a more effective counter to the complications from perianal Crohn's.

MCQs

1) Select ONE statement below that applies to ulcerative colitis:

a) It can affect any part of the alimentary system from mouth to anus.
b) Inflammation is diffuse but limited to the colonic mucosa.
c) Inflammation is transmural but patchy and segmental.
d) Smoking is shown to predispose this disease.

2) Which criteria is used to classify the severity of ulcerative colitis to guide medical management? Select ONE.

a) APACHE II
b) Glasgow-Ranson
c) Truelove and Witts
d) Rockall

3) The following are types of pouches for ileo-anal pouch procedures (IAPP), EXCEPT,

a) Y-pouch
b) J-pouch
c) S-pouch
d) W-pouch

4) Below are the potential indications of stricturoplasties in small bowel strictures, EXCEPT,

a) Multiple strictures.
b) Patients with small bowel syndrome
c) Strictures without associated phlegmon or fistula
d) Perforated bowel

5) Select the appropriate techniques from below that are stricturoplasties for small bowel strictures

a) Heinecke-Mikulicz
b) Finney
c) Billroth
d) Whipples
e) Ellis
f) Side-to-side isoperistaltic
g) Side-to-end isoperistaltic

Answers

1) b
2) c
3) a
4) d
5) a,b and f

INFLAMMATORY BOWEL DISEASE: SURGICAL PERSPECTIVES

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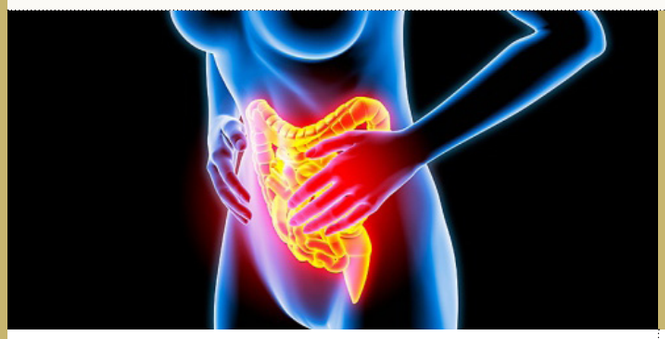

Inflammatory bowel disease: surgical perspectives
General Surgery

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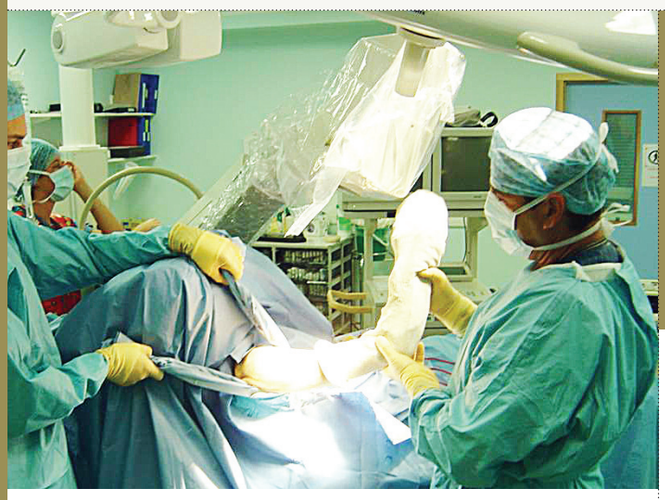
PROXIMAL HUMERUS FRACTURES IN THE ELDERLY

DLJ Morris, C Quah



Abstract

Proximal humerus fractures in the elderly are common and frequently result from a fall onto an outstretched hand. Management is determined by assessment of the patient and fracture. We discuss the diagnosis, classification and management options for these injuries.

Keywords: *Proximal humerus fractures, elderly, classification, management, education.*

Case

A 61 year old lady sustained a closed 4-part displaced proximal humerus fracture following a fall onto her left outstretched hand. The injured arm was initially immobilised a sling. Following a thorough assessment of the patient a left shoulder hemiarthroplasty was performed.

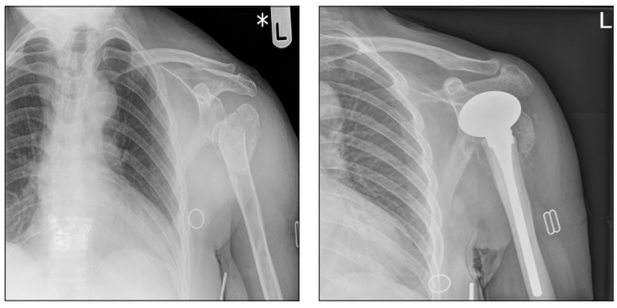


Figure 1: A 4-part proximal humerus fracture treated with a shoulder hemiarthroplasty.

Introduction

Fractures of the proximal humerus are relatively common, representing 4-5% of all fractures. Incidence increases with age with the highest incidence in women between 80 and 89 years of age. (1) A variety of management options are available. Given this, debate exists regarding management of proximal humerus fractures in the elderly. This article discusses clinical assessment of an elderly patient presenting with a proximal humerus fracture, the Neer 4-part classification system and management options.

Diagnostic Considerations

History and examination

History and examination is critical to the identification of proximal humerus fractures, particularly in the elderly. 87% of fractures in adults result from falls onto an outstretched hand from a standing height. The proximal humerus is a common site for such a fracture to occur in the elderly, with women affected more than twice as frequently as men.

History should include the mechanism of injury and circumstances precipitating this. Though commonly a consequence of a mechanical fall, it is important to exclude an underlying medical cause for the fall in the elderly, such as syncope. The mechanism of injury will also guide the clinicians index of suspicion for associated injuries.

A thorough past medical and drug history should be obtained. This should include medical comorbidities and level of function. Tobacco use should be documented. It is particularly important to establish the level of function the patient had prior to the injury in the affected shoulder. Pre-existing rotator cuff pathology or significant glenohumeral osteoarthritis may influence management options. The presence of osteoporosis is also significant as patients with osteoporotic proximal humerus fractures have more complex fractures. (2)

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Patient risk factors for a proximal humerus fracture include poor quality bone, impaired vision and balance, medical comorbidities and decreased muscle tone.

Clinical examination should initially focus upon identifying a precipitant for the fall, such as sepsis, and excluding significant associated injuries. Rib, scapula and cervical fractures are commonly associated with proximal humerus fractures in the elderly. After this a thorough assessment of the affected arm should be undertaken. It should be established whether there is an open fracture, and a neurovascular assessment should be performed. The brachial or radial artery should be palpated and capillary refill time of the fingers assessed. Particular attention should be paid to the axillary nerve by testing for sensation in the regimental badge area over the deltoid muscle, and deltoid power. 21-36% of proximal humeral fractures produce neurovascular injury. (3) 5% have a concurrent brachial plexus injury. Ecchymosis usually occurs at the fracture site within 24-48 hours of injury.

Imaging

Investigations should include anteroposterior and scapular lateral x-ray views of the affected humerus. Axillary views can be used at assess the humeral head and gleno-humeral articulation but have a high risk of causing fracture displacement and should ideally be avoided. CT imaging should be used in more complex cases to assess comminution or posteriorly displaced greater tuberosity or medially displaced lesser tuberosity fragments. MRI may be used to assess rotator cuff pathology.

Classification

The most frequently used system for classification of proximal humeral fractures is Neer's 4-part system based on the 4 usual cleavage lines that occur due to the anatomy of the proximal head of the humerus. (4)

Neer considered there to be four segments of the proximal humerus - the articular part, the greater tuberosity, the lesser tuberosity and the humeral shaft. Two-part, three-part and four-part fractures can occur. The fractures are then classified by their degree of displacement and angulation. Surgical neck fractures are the most common type of proximal humeral fracture. These lie just below the greater and lesser tuberosities. 3 and 4-part fractures represent 13-16% of proximal humerus fractures.

The tuberosities are insertion sites for the rotator cuff, with supraspinatus, infraspinatus and teres minor inserting into the greater tuberosity and subscapularis inserting into the lesser tuberosity. Therefore fractures involving these insertion sites can become displaced due to the pull of the rotator cuff muscles. The humeral shaft is also pulled medially by pectoralis major following a proximal humerus fracture.

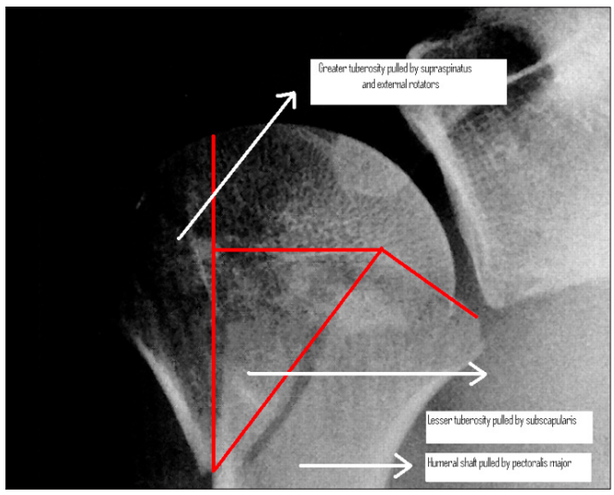

Figure 2: Labelled radiograph displaying the displacing forces creating by the pectoralis major and rotator cuff muscles in a 4-part proximal humerus fracture.

Management Considerations

Initial management includes analgesia and immediate immobilisation. Associated dislocation should be reduced. Open fractures should prompt urgent senior review and should be managed according to BOA/BAPRAS guidelines.

Conservative treatment is preferred for non- or minimally displaced fractures (5) particularly among the elderly given that fracture fragments are generally held together by muscles, tendons, rotator cuff attachments and the periosteum. Most of these proximal humerus fractures will heal without surgery, and many recover satisfactory function following use of a supportive sling and early rehabilitation. 2-part non-displaced fractures are the most common variant.

Conservative management is indicated for greater tuberosity fractures with less than 5mm of posterior or 10mm of superior displacement in active patients. The greater tuberosity fragment is pulled superiorly by the supraspinatus and posteriorly by infraspinatus and teres minor. Surgical neck fractures with any bony contact between fragments are managed conservatively in the elderly.

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Sling immobilisation should occur for 7-10 days with active finger, wrist and elbow movements encouraged. Gentle, active, assisted range of movement exercises should start 2 weeks post injury, with light resistance exercises introduced 4 weeks after. At 3 months patients should aim to commence shoulder strengthening exercises.

Debate continues regarding treatment of displaced fractures and indications for surgical management. Displacement is defined as 1 cm between fragments or 45° of angulation between fracture fragments.

Considerations for surgical management include fracture type, bone quality, assessment of rotator cuff and patient age, activity level and comorbidities.

Surgical management of displaced 2-part fractures

Indications for surgical intervention for 2-part fractures include open fractures, inability to obtain or maintain a closed reduction and injury to the axillary artery. 2-part fractures can be managed with percutaneous K-wire fixation if there is not significant comminution or open reduction and internal fixation with a proximal humeral internal locking system (Philos) plate.

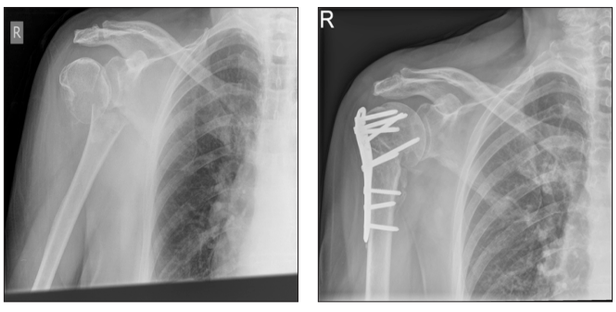

Figure 3: A 2-part proximal humerus fracture treated with a Philos plate.

Displaced greater tuberosity fractures retract posteriorly and superiorly, making a closed reduction difficult. If left in position, impingement will develop against the acromion, limiting elevation and external rotation of the shoulder. Therefore significantly displaced greater tuberosity fractures should be managed surgically alongside repair of any associated rotator cuff tears. Additional fixation methods for greater tuberosity fractures include non absorbable sutures and cancellous lag screws.

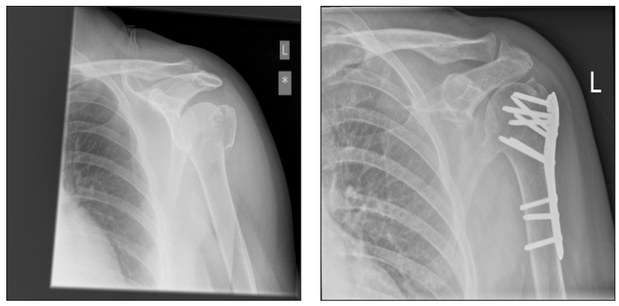

Figure 4: A displaced greater tuberosity fracture treated with a Philos plate.

Surgical management of displaced 3-part fractures

3-part fractures can be managed with open reduction and internal fixation using a Philos plate. In these fractures one of the tuberosities remains with the articular head fragment, thereby retaining its vascularity.

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Surgical management of displaced 4-part fractures

These injuries commonly cause avascularity of the articular segment which even with satisfactory reduction and fixation would eventually collapse. Management options include a shoulder hemiarthroplasty or reverse total shoulder replacement. Indications for hemiarthroplasty in 4-part fractures include dislocation, medial metaphyseal comminution, and lateral head displacement. Problems associated with hemiarthroplasty include tuberosity displacement, prosthesis malpositioning, functional limitations and poor rehabilitation.

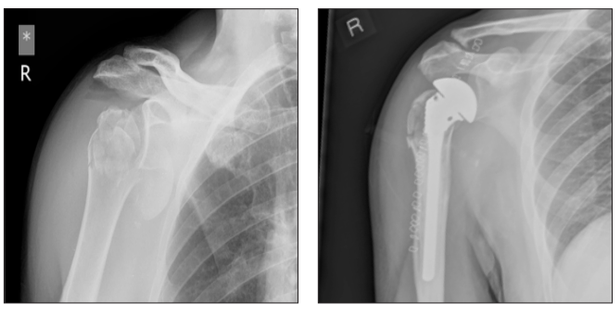

Figure 5: A 4-part proximal humerus fracture treated with a shoulder hemiarthroplasty

Reverse total shoulder replacement should be considered for patients older than 75, for women with osteopenia and for patients with comorbidities, poor blood supply and poor observance of rehabilitation. Problems associated with reverse total shoulder replacement include loss of external rotation, instability, infection, and implant deterioration and loosening.

Additionally, 2-part displaced anatomical neck fractures often render the articular surface avascular and require prosthetic replacement with a shoulder hemiarthroplasty.

Reverse total shoulder replacement is a recently introduced management option, with shoulder hemiarthroplasty traditionally being the treatment of choice for 4-part fractures. However, Bufguin et al (6) published a randomised study in 2007 comparing reverse total shoulder replacement and shoulder hemiarthoplasty in acute fracture patients. Each group comprised 28 patients older than age 70. At 33 months follow-up reverse total shoulder replacement restored anterior active elevation better than shoulder hemiarthroplasty (122 degrees vs. 109 degrees, p = 0.07) and reverse total shoulder replacement was slightly better than shoulder hemiarthroplasty in relieving pain. External rotation, however, was better in the shoulder hemiarthroplasty group.

Outcomes

Prognosis depends on fracture type, mechanism of injury and the patient's age and underlying health. Complications include avascular necrosis, nerve injury, malunion, non union and rotator cuff dysfunction. A lesser tuberosity non union leads to weakness of subscapularis. A greater tuberosity non union leads to a lack of active shoulder elevation. In general, an elderly person with a proximal humeral fracture never regains full range of movement, whether treated conservatively or surgically. The aim is rehabilitation to a functional range of movement. Generally recovery takes at least 1 year but union is expected at 6-8 weeks.

Discussion

A recent Cochrane review found there is insufficient evidence to inform the management of proximal humerus fractures, particularly to establish what is the best method of surgical treatment, either in terms of the use of different categories of surgical intervention or different methods of performing an intervention in the same category (such as different methods of plate fixation). (7)

Only 12 small randomised trials with 578 participants were identified. One trial suggested hemiarthroplasty resulted in better short term function with less pain and disability in comparison to conservative management in severe injuries, but other trials highlighted that conservative management had comparable and often favourable outcomes in comparison to surgical management. The review highlights a need for 'good quality evidence for the management of proximal humerus fractures'.

The Proximal Fractures of the Humerus: Evaluation by Randomisation (ProTHER) is a multicentre randomised controlled trial currently being undertaken in the United Kingdom. It aims to evaluate the effectiveness and cost-effectiveness of surgical versus non-surgical treatment for the majority of displaced fractures of the proximal humerus in adults. Participants are patients with a radiologically confirmed displaced fracture of the proximal humerus with involvement of the surgical neck with randomisation to surgical and non-surgical groups. The trial should address the current lack of 'good quality evidence' in relation to management of proximal humerus fractures. The results of this trial will be presented next year at the British Elbow & Shoulder Society conference. (8)

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Conclusion

Proximal humerus fractures in the elderly are classified using the Neer 4-part classification and management depends upon fracture pattern, degree of displacement and angulation and patient factors. Conservative management is often preferred in the elderly population, particularly in undisplaced fractures. Surgical options are dependent upon fracture pattern, with K-wire fixation, Philos plate, shoulder hemiarthroplasty and reverse total shoulder replacement all utilised when appropriate. Poor evidence exists to support current management of proximal humerus fractures.

MCQs

1. Which fracture is the most common type of proximal humeral fracture in the elderly?

a. Anatomical neck

b. Surgical neck

c. Greater tuberosity

d. Lesser tuberosity

2. What mechanism of injury typically causes a proximal humeral fracture in the elderly?

a. Direct blow to arm

b. Road traffic accident

c. Fall onto outstretched hand

d. Epileptic seizure

3. Which of the following is a surgical treatment for a proximal humeral fracture in the elderly?

a. Hemiarthroplasty

b. Plate fixation

c. Reverse total shoulder replacement

d. All of the above

4. Which of the following is not a segment in Neer's 4-part classification?

a. Greater tuberosity

b. Lesser tuberosity

c. Humeral shaft

d. Intertubercular groove

5. Which of the following is not a recognised complication of a proximal humerus fracture in the elderly?

a. Malunion
b. Avascular necrosis
c. Axillary nerve injury
d. Frozen shoulder

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Answers

1. b

2. c

3. d

4. d

5. d

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Proximal humerus fractures in the elderly
Trauma & Orthopaedic Surgery

Disclaimers

CRANIOSYNOSTOSIS - A GUIDE

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Abstract

Craniosynostosis is a common craniofacial condition that can affect 1 in 2500 people. It can be divided into single-suture, multi-suture and syndromic. Non-syndromic is more common than syndromic but the management remains the same in surgical correction of the suture. This article discusses craniosynostosis and its management.

Keywords: *Craniosynostosis, craniofacial, syndromic.*

Case Study

A three month old baby boy accompanied by the parents attends their local paediatric unit following concerns about the baby's head shape and circumference. The paediatrician refers them to their regional Craniofacial Unit where following a history, examination and 3D-CT scan; it is found that the left sagittal suture is fused whereas the other sutures are normal. The baby has the pi procedure to allow for cranial expansion. The recovery was uneventful.

Introduction

Craniosynostosis is defined as the premature fusion of the cranial sutures and has an incidence of 1 in 2500 (1). In 1851, Virchow created a classification system for the types of skull deformity observed in craniosynostosis and made the observation that premature fusion results in compensatory growth in other areas of the skull (2). Today, craniosynostosis is defined as involving a single suture versus multiple sutures and as either syndromic or non-syndromic.

Diagnosis

The diagnosis of craniosynostosis begins with a visual examination for signs of dysmorphism. Deviations in the shape of the cranial vault, size and position of the orbits, projection of the midface, side-to-side asymmetries and tissue deficiencies would lead a clinician to refer to an established craniofacial unit. Examination of the extremities (table 1) may reveal the presence of syndactyly of the second, third and fourth fingers (Apert syndrome), broad distal phalanges in the thumbs and great toes (Pfeiffer syndrome) and so help guide the diagnosis (3).

The usefulness of routine CT scanning has been questioned in the management of non-syndromic craniosynostosis, imaging is a necessary part of the investigative process. 3D-CT scanning provides important information about bony architecture, suture pattern, brain anomalies and venous drainage anomalies (3). However, its use remains controversial for single-suture synostosis (4). MRI imaging is a useful adjunct for brain analysis and dental radiographs such as OPG may be useful in assessing dental anomalies. It is also important to perform ophthalmological screening as there are often visual abnormalities accompanying craniosynostosis (3).

Craniosynostosis	Thumbs	Hands	Great toes	Feet
Apert	Occasionally fused to fingers	Complex acrocephalosyndactyly	Occasionally fused to toes	Complex syndactyly
Crouzon	Normal	Normal	Normal	Normal
Muenke	Normal	With/without carpal fusion	Broad	With/without tarsal fusion
Pfeiffer	Broad, medially deviated	Variable brachydactyly	Broad, medially deviated	Variable brachydactyly
Saethre-Chotzen	Normal	Variable syndactyly	Broad/duplicated	Variable syndactyly

Table 1: Distinguishing extremity features in craniosynostosis.

Non-syndromic craniosynostosis

The non-syndromic subtype is present in 0.4-1 in 1000 births (5) and unlike the syndromic craniosynostoses there are no other dysmorphisms of the face, trunk and extremities (5). Furthermore, non-syndromic craniosynostoses usually involve a single suture, the most common types being sagittal, unicoronal, bicoronal, metopic and lambdoidal. Although rare, multiple suture synostosis does exist and is referred to as complex. Sagittal synostosis is the most common form of craniosynostosis and comprises 45% of non-syndromic cases (6). Sagittal synostosis leads to a boat-shaped deformity of the skull (scaphocephaly) with growth restriction in width and compensatory growth in calvarial length in the anterior to posterior direction. This can result in varying degrees of frontal bossing and occipital coning (6). Sagittal synostosis has a male-to-female ratio of 4:1(1).

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Unicoronal synostosis is involved in 25% of non-syndromic cases (6). It consists of anterior plagiocephaly with ipsilateral flattening of the forehead on the affected side and contralateral bulging of the frontoparietal skull (1). This growth restriction results in a facial twist that arises from compensatory pressure of the ipsilateral temporal lobe pushes the maxilla forward causing a rotation of the midface.

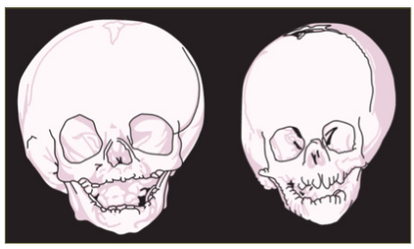

Figure 1: Unicoronal synostosis.

There is deviation of the ipsilateral zygoma forward as well as rotation of the maxilla that the nasal tip is deviated to the contralateral side. Bilateral coronal fusion causes brachycephaly, skull shortening in the anterior to posterior dimension and turricephaly, skull lengthening in a cranial to caudal direction (7). Unicoronal synostosis is more common in females than males by 3:2. Metopic synostosis occurs in 25% of non-syndromic cases (6).

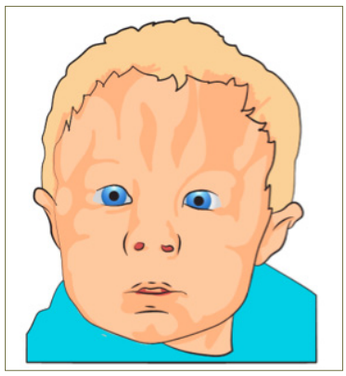

Figure 2: Metopic synostosis.

Trigonocephaly is characterised by a triangular-shaped forehead with bifrontal and bitemporal narrowing and parietal and occipital prominence. This leads to the appearance of hypotelorism and a low nasal dorsum with epicanthal folds. The rarest type is lambdoidal synostosis which is characterised by occipital dysmorphism.

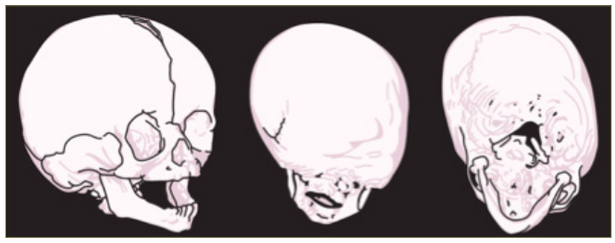

Figure 3: Right lambdoid synostosis.

The result is an ipsilateral mastoid bulge, thickened ridging of the affected suture and tilt of the occipital skull base with the affected side shifted downward. Inferior displacement of the skull base pulls the external auditory canal downwards on affected side, leading to inferior displacement of the entire ear. This can be useful in differentiating between positional plagiocephaly and lambdoidal synostosis (8). Constriction in the lambdoid region causes a compensatory bulge in the contralateral posterior parietal region (8).

Syndromic craniosynostosis

Apert syndrome

This condition was first described in 1894 and is characterised by turribrachycephaly, midface hypoplasia and symmetric syndactyly of both hands and feet. It is the result of mutations of FGFR-2 that occurs in 1 in 100,000 births and although has an autosomal dominant inheritance pattern, the majority of cases seen have sporadic mutations (9). The most common presentation is bicoronal synostosis with a characteristically large anterior fontanelle, bitemporal widening and occipital flattening.

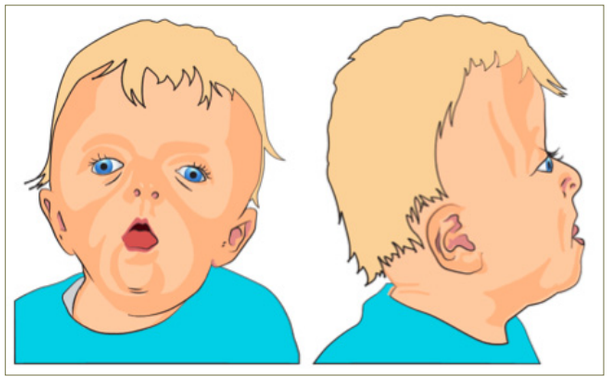

Figure 4: Apert syndrome.

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These patients also have midface hypoplasia with concavity of the midface, shallow orbits with mild hypertelorism and downward slanting palpebral fissures, ocular proptosis, a high arched or cleft palate, anterior open bite. They also have a depressed nasal bridge and downward turned tip (9).

The hand syndactyly which is pathognomonic for the condition, often involves the second, third and fourth fingers leading to a middigital hand mass. In some cases, the first and fifth fingers join the middigital mass. If the thumb is free, it is broad and deviates radially. In the feet, the syndactyly involves the second, third and fourth toes. These patients have an increased incidence of delayed mental development and 70% will have acne vulgaris (9).

Crouzon syndrome

This condition is characterised by brachycephaly, shallow orbits with ocular proptosis, midface hypoplasia and an anterior open bite.

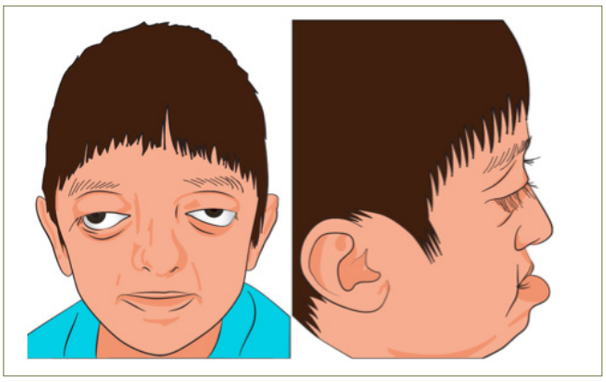

Figure 5: Crouzon syndrome.

The defining characteristic that separates Crouzon syndrome from Apert syndrome is that there are no limb anomalies (9). It is caused by mutations of FGFR-2 and demonstrates an autosomal dominant inheritance pattern. It is observed in 1 in 25,000 births making this the most common of the syndromic craniosynostoses. These patients demonstrate a spectrum of phenotypic variability. One common scenario that presents is where a mildly affected parent and previously undiagnosed parent have a more severely affected child (9).

Bicoronal synostosis is the most common pattern leading to a brachycephalic head although scaphocephaly, trigonocephaly and the cloverleaf skull deformity have been described in this condition. Fusion of the cranial base sutures lead to shallow orbits with exorbitism. Midface hypoplasia together with a constricted high arched palate and an anterior open bite are features. These patients typically have normal intelligence (9).

Muenke syndrome

This was first described as a result of the identification of the genetic mutation as opposed to the phenotype. The mutation is a pro250Arg mutation in FGFR-3 on chromosome 4p31 (10). The incidence is 1 in 10,000 and demonstrates an autosomal dominant inheritance pattern with variable expressivity (11). It has been estimated that Muenke syndrome may be present in 10% of unicoronal and bicoronal synostosis cases that were thought to be non-syndromic (12). The general features are craniosynostosis of the coronal sutures, hearing loss, developmental delay and thimble-like middle phalanges.

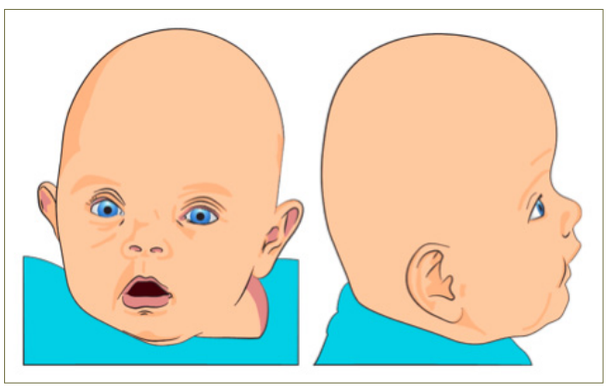

Figure 6: Muenke syndrome.

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Midface hypoplasia is an uncommon finding. There is variability in the presentation between genders. 88% of females and 76% of males have the mutation have craniosynostosis (12). Bicoronal synostosis is the most common presentation but in males, there is a higher incidence of unicoronal synostosis (29% compared to 20% of females) (11).

Pfeiffer syndrome

This syndrome consists of a spectrum of craniofacial features ranging from mild to severe. The most common features include: turribrachycephaly, midface hypoplasia, exorbitism, and broad thumbs and great toes and variable soft tissue syndactyly. Other features include: hyperterlorism, strabismus, downward slanting palpebral fissures, a beaked nasal deformity and class III malocclusion. The majority of cases of Pfeiffer syndrome involve FGFR-2 mutations but 5% of mutations affect FGFR-1 and demonstrate a less severe phenotype (13). The incidence is 1 in 100,000 births and the inheritance pattern is autosomal dominant.

Saethre-Chotzen syndrome

It has an incidence of 1 in 25,000 to 1 in 50,000 births and is characterised by a range of phenotypes and severity caused by the autosomal dominant inheritance of mutations of the TWIST-1 gene on chromosome 7p21.2 (14). The variability of presentation is highlighted by the heterogeneous pattern of craniosynostosis. The majority of patients present with bicoronal synostosis (45-76%) but unicoronal (18-27%) and other multi-suture presentations are common (15). Other features include: a low frontal hairline, eyelid ptosis, and facial asymmetry plus ear deformities with a characteristic prominent crus helicis extending through the conchal bowl (16). A minority of these patients will have hearing loss, brachydactyly, syndactyly and clinodactyly (17). Midface hypoplasia is not a common feature of Saethre-Chotzen syndrome.

Treatment

Non-syndromic craniosynostosis

The general indications for surgical intervention in non-syndromic craniosynostosis include the presence of cosmetic deformity and/or functional impairment such as an elevated intracranial pressure (ICP) (18). The majority of surgeons delay procedures until the patient is 3 months old and is able to withstand the insults of surgery such as bleeding. A popular argument for early intervention is minimising cerebral constriction (19). However, it has been shown that intracranial volume normalises by 6 months. Therefore, surgical correction to allow for cranial expansion should be performed in the first few months. There is no direct correlation between intracranial pressure and intracranial volume and also no link between intracranial volume and neurocognitive impairment exists, so this weakens the argument (20).

There is no consensus on the optimal timing of surgical correction. The timing is influenced by surgeon preference, timing of referral to the appropriate specialist and the surgical technique to be used (table 2). If you use an endoscopic procedure, it is best that it is performed as early as possible (< 3 months). Open techniques do not have such rigid age limits. It is also important to be aware that there is no exact consensus on the surgical techniques used for skull reconstruction. There are many techniques that have been described but those presented here are commonly used (20).
Sagittal synostosis

Synostosis	Procedure(s)
Sagittal	Synostectomy (open/endoscopic) Pi procedure Cranial vault reconstruction
Coronal	Frontoorbital advancement
Metopic	Frontoorbital advancement
Lambdoidal	Bilateral occipital and parietal reconstruction

Table 2: The surgical management of non-syndromic craniosynostosis.

There are a range of approaches available for managing sagittal synostosis. One example is synostectomy which can be performed open or endoscopic or cranial vault reconstruction (21). However, one such approach is the pi procedure. Greater degrees of deformity will require lateral wedge, radial/ frontal and occipital osteotomies and subtotal calvarial reconstruction. For more severe cases and for children presenting later, a two stage procedure may be considered in which occipital deformity is corrected at stage one and stage two will be frontal reconstruction (22).

Coronal synostosis

Correction requires a frontal reconstruction that addresses the superior and lateral periorbital skeleton as well as the forehead. Most commonly performed is the fronto-orbital advancement. A bifrontal craniotomy must be performed that can be done as a single piece or a double piece. In unilateral cases, the frontal bandeau is cut to bring the lateral orbital rim on the affected side (20). It is believed that this prevents a bony step-off on the zygomatico-frontal buttress after the advancement is performed. The bandeau is weakened in the glabellar region in order to bring the affected side forward (20). There are three pivot points: one is located at the unaffected zygomatico-frontal suture, the second is in the glabellar region and the third is at the body of the zygoma at the affected side. They move in different directions and allow for three dimensional changes that result from advancement of the bandeau. The glabellar point is stabilised with plates and screws mainly on the internal side. The lateral temporal wing of the bandeau is made longer on the affected side (20).

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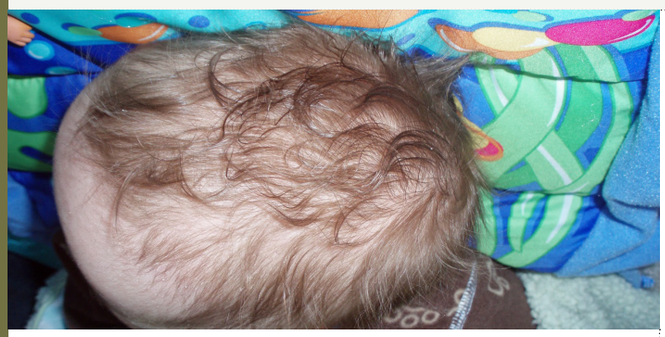

Metopic synostosis

It is important that this is distinguished from a normally fused metopic ridge. Patients with metopic synostosis are born with a fused suture and trigonocephaly. They also have more medial orbits which make them look more hypoteloric (23). Surgical correction requires frontal reconstruction that addresses the superior and lateral periorbital skeleton and forehead. This is done between 8 to 12 months of age. An open approach can be used that allows for complete fronto-orbital advancement. A bifrontal craniotomy is performed and the frontal bandeau is removed with bilateral temporal extensions. The lateral orbital rims are also included in the bandeau. Trigonocephaly can result in a narrow frontal bandeau so it is important to widen by splitting it in the midline and inserting a bone graft fixed into place with resorbable plates and screws. Once fixed in place, twisting is performed to optimise brow projection. The frontal bone may also be too narrow, so contouring alone may be insufficient to achieve full alignment. So split the bandeau and contour as necessary to place it back as two separate pieces. Barrel stave osteotomies can also be performed in the parietal bone behind the coronal sutures to widen the vault (20).

Lambdoidal synostosis

Correction of either bilateral or unilateral synostosis requires bilateral occipital and parietal reconstruction. Posterior vault reconstruction is performed between 3-6 months. Bilateral posterior parietal-occipital craniotomies are made. The anterior extent of the parietal cuts is made anterior to the compensatory bulging. The entire posterior parietal-occipital bone can be removed as a single piece or two pieces with an occipital bandeau (20). The occipital bandeau is contoured and flipped to increase the volume on the constricted side. The posterior parietal bones are also flipped and orientated to achieve the most normal skull contour possible. Rigid internal fixation is achieved with resorbable plates and screws. Surgical cranial defects will be present and can be left to reossify (20).

Syndromic craniosynostosis

Different surgical procedures are performed depending on the age and requirements of the patients (table 3). The goals of surgical treatment in the first year of life are to increase the intracranial volume so that the risk of developing elevated intracranial pressure (ICP) is reduced and improving head shape. The techniques most commonly used for the initial vault expansion are; fronto-orbital advancement with anterior cranial vault remodelling or posterior vault expansion (24). These procedures can be performed as one stage techniques.

However, other techniques such as cranial vault distraction and spring assisted cranioplasty are being used as they can augment the change in head shape and increase intracranial volume with a reduced morbidity. However, the timing for these procedures can be controversial and is largely dictated by changes in the physical properties of the calvarium in infancy.

Before 6 months, the cranial bones are malleable but too weak to support rigid fixation. After 12 months, the bones are thicker and more rigid so fixation is more effective but there is a reduction in malleability. Also it is important to be aware that the innate ability of the infant skull to spontaneously heal large bony defects is lost between 9 and 11 months (25). Therefore, the general consensus is that open vault remodelling should take place between 6-9 months. If there is confirmed or suspected elevated ICP, urgent decompression with strip craniectomy needs to be performed if the patient is less than 3 months, spring assisted cranioplasty between 3-6 months or a distraction technique if the patient is over 6 months.

Age (years)	Procedure(s)
< 1	Frontoorbital advancement (FOA) Posterior vault distraction Spring assisted cranioplasty
Mid childhood	Midfacial advancement (Monobloc or distraction osteogenesis)
Adolescence	Orthognathic surgery Final facial contouring

Table 3: Timing for surgical procedures for patients with syndromic craniosynostosis.

Posterior vault distraction is advantageous in patients with severe turricephaly and occipital flattening as it allows for an expansion of the intracranial volume and improvement in head shape (26).

Surgery during childhood

The timing of midface correction is controversial. Some centres advocate early correction whereas others will wait until skeletal maturity has been reached. There are advantages and disadvantages to earlier intervention or delayed intervention. Midface advancement is achieved by performing Le Fort III subcranial osteotomies. This leads to craniofacial disjunction and midface advancement. This can be done in either a single stage (27) or by distraction osteogenesis (28). The midface can be advanced alone or in combination with the supraorbital bar and frontal bone flaps as a monobloc procedure. When the correct supraorbital rim to cornea relationships is present, the monobloc procedure allows for simultaneous correction of the brow, supraorbital and midface deformities (29).

However, there are complications with this procedure such as significant blood loss, CSF leak and frontal bone flap necrosis (30). To reduce the risk of CSF leak, an alternative intrasinus approach has been suggested (31).

Regardless of the timing of midface advancement, distraction has become the treatment of choice for the extensive midface advancement these patients require. Distraction allows for expansion of the soft tissue envelope with the bony advancement, reducing relapse from soft tissue contraction.

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The goals of midface distraction is tailored to each patients deformity, the universal goals are maximal projection of the zygoma and restoration of orbital volume. This results in a transition from a severe class III malocclusion to a class II malocclusion. The final occlusion can be addressed with orthodontic management and orthognathic surgery once skeletal maturity has been reached. There are advantages and disadvantages to distraction (table 4).

Overall, distraction osteogenesis has significantly improved the results obtainable for midface advancement whilst reducing the complications (32-33). Increasingly, other alternatives are being attempted such as performing Le Fort II midface distraction and simultaneous zygomatic repositioning. The major criticism of Le Fort III distraction is that it advances the midface but in conditions such as Apert syndrome, the central concavity and vertical compression remain untreated. One study showed that with Le Fort II segmental movement, there was increased length and advancement of the central face compared to the lateral orbit (34). The authors commented that further study is required.

Advantages	Disadvantages
Less blood loss	Prolonged time needed for distraction and consolidation
Greater advancement (up to 20 mm)	Second procedure required to remove buried devices
Elimination of bone grafts	Requirement of an external halo device for a prolonged period
Reduced infection risk	
Less relapse	

Table 4: The advantages and disadvantages of distraction.

Surgery at adolescence

The abnormal pattern of facial growth in children with syndromic craniosynostosis often leads to dentofacial deformities. Class III malocclusion, secondary to midface hypoplasia is the most common deformity and often develops despite midface surgical treatment (28). The approach is team support between the orthodontist, dentist (if needed) and the surgeon. Following completion of maxilla and mandibular growth, surgery may be required. This consists of orthodontic management to optimise the bite for at least 18 months followed by osteotomy. After surgery, orthodontic management will continue for another 6 months. The surgical procedures will be performed once maturity of the facial skeleton has been reached (35).

Surgical complications and outcomes

There are acute and chronic complications following open surgery for craniosynostosis (table 5). The acute complications include: bleeding, infection, CSF leak and stroke (1). The chronic complications can include: failure of re-ossification and contour irregularity leading to a need for a repeat procedure. One study assessed all their cases of craniosynostosis over a 12 year period. They found that compared to a previous study performed in 2002 there was an improvement in blood loss, operative time and average length of hospital stay. They achieved a lower complication rate of 3.3%, no deaths and a reoperation rate of 10.8%.

It was also shown that those who underwent surgical correction before the age of 6 months were had higher complication rate and higher rates of re-operation (36).

In a cohort study of 3000 patients that underwent surgical repair, the overall mortality rate was <1%, the complication rate of 10% and a mean length of stay of 4.2 days. They showed that patients aged 1-3 years at the time of surgical intervention were more likely to have a longer length of stay which they also argued could be a marker for a higher complication rate.

In a separate study, a team investigated the long-term outcome of unilateral coronal synostosis. It was found that osseous dysmorphology did not normalise and there was a tendency for reversion towards the untreated phenotype by the time skeletal maturity was reached. This was confirmed in another study that showed that despite normalised cranial indices, postoperative growth was limited in all types except lambdoidal synostosis. A correlation has also been identified between the severity of postoperative growth restriction and earlier age at the time of surgical intervention (37).

Acute	Chronic
Bleeding	Failure of reossification
Infection	Contour irregularity
CSF leak	Reversion to untreated phenotype
Meningitis	
Stroke	
Death	

Table 5: Surgical complications of repair of craniosynostosis.

Conclusion

Craniosynostosis is commonly encountered in craniofacial surgery. It has been found that surgery is needed for both cosmetic and functional reasons. Non-syndromic craniosynostosis is more common but still need multidisciplinary support along with their syndromic counterparts. The ideal place of care is a tertiary craniofacial unit where multidisciplinary support is available.

After studying this article you should be able to:

1. Understand syndromic and non-syndromic craniosynostoses and be able to identify them when examining a patient.
2. Be able to perform correct investigations for a patient with suspected craniosynostosis.
3. Be able to explain the surgical management of syndromic and non-syndromic craniosynostosis.

Questions

1. What is the incidence of craniosynostosis?

a) 1 in 1000
b) 1 in 2500
c) 1 in 5000
d) 1 in 10,000

2. What is the most common non-syndromic synostosis?

a) Coronal
b) Lambdoid
c) Sagittal
d) Metopic

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3. What helps to differentiate Apert syndrome from Crouzon syndrome?
a) Midface hypoplasia
b) Cleft palate
c) Bicoronal synostosis
d) Syndactyly of the fingers and toes

4. What is a significant problem for some syndromic craniosynostosis patients?
a) Raised ICP
b) Strabismus
c) Abnormal GI function
d) Endocrine abnormalities

5. What are the general indications of surgical intervention in non-syndromic craniosynostosis?
a) Cosmetic deformity
b) Functional impairment
c) Parental pressure
d) Hospital policy

Answers

1. (b) 2. (c) 3. (d) 4. (a) 5. (a or b)

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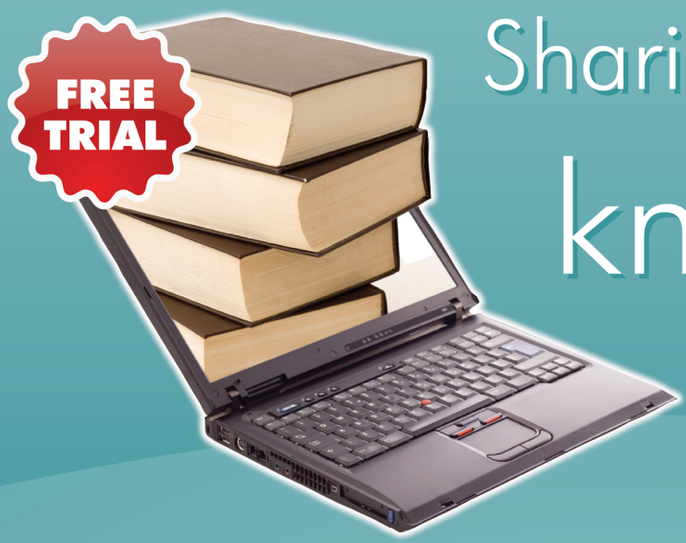

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PERICARDIAL EFFUSIONS & COLLECTIONS AFTER CARDIAC SURGERY

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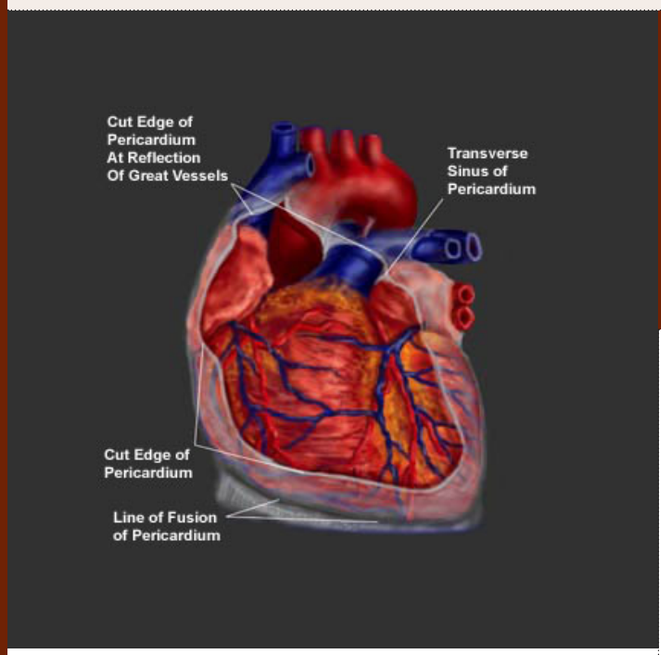

Abstract

Pericardial collections/effusions can occur at any time during the post operative period. After cardiac surgery the aetiology of these collections may vary depending on the timing of the collection. These can be divided into immediate (<24 hours) or delayed presentations, within which onset could be days or even months. Post-operative bleeding contributes to a significant amount of collections, this can be immediately post operatively, days, weeks or even months after the operation. Initially bleeding may be due to complications from the procedure, whereas anticoagulation post-operatively plays a greater role in delayed pericardial effusions. Other causes include heart failure, postpericardiotomy syndrome and chylopericardium. The investigation of these collections does not vary greatly, with echocardiography used as the gold standard. However more complex investigations (CT, MRI) may be useful in difficult cases. Management of pericardial collections involves early resuscitation as required followed by more targeted treatment taking into account several factors such as the size, location, local effects of the collection as well as the patient's general state.

Keywords: *Pericardial, effusion, collection, postpericardiotomy, tamponade.*

Introduction

Pericardial effusions are noted to occur in the vast majority of patients after cardiac surgery. Incidence is quoted from 1% to 85% post operatively, the variance largely attributable to the study definitions and design. The general incidence of pericardial effusions when studied prospectively with frequent echocardiographic examinations is noted to be as high as 64% (1), 85% (2) and 77% (3). These varied in size and significance. In contrast, retrospective studies looking at moderate and large effusions have shown an incidence of 1-2% in cardiac surgery patients (4,5).

Pericardial effusions range from very small to very large and there is no relationship between the size and subsequent development of cardiac tamponade. Cardiac tamponade is defined as fluid within the pericardial sac resulting in increased intrapericardial pressures impairing the hearts ability to fill and pump. Cardiac tamponade is occurs in 0.1% to 6% of cardiac surgery patients (6,7).

Postoperative pericardial effusions may occur anytime from the immediate postoperative period to months after the surgery. Generally effusions reach their maximum size by day 10 post operatively and regress spontaneously thereafter (2).

Pericardial effusions are associated more with valvular surgery, especially aortic valve replacement (4). The incidence is lower in patients who undergo coronary artery bypass grafts (CABG) due to the free connection between the mediastinum and the pleural space which is opened for harvest of the left internal mammary artery. Furthermore, CABG patients did not undergo routine echocardiograms unless clinically indicated thereby reducing pericardial effusion pick up rates. Patients undergoing aortic root surgery (4,15,16) are at an increased risk. This may be due to greater early postoperative bleeding in these patients, inflammation secondary to retained mediastinal clot, from osmotic fluid accumulation in the peri-graft space after a clot lysis and disruption of lymphatics due to significant medisastinal dissection required for these operations. Association between renal failure and pericardial effusions post operatively has been noted, supporting the argument for serosal fluid accumulation in fluid retention.

Immediate postoperative pericardial effusions are most frequently due to bleeding. Those occuring later in the post operative period are noted to be linked to bleeding, heart failure, chylopericardium and post pericardiotomy syndrome.

Ashikhmina et al (4) noted that pre-operative anticoagulant use and preoperative haematocrit are also related to later onset pericardial effusions. They also noted that delayed pericardial effusions were more likely in patients undergoing heart transplants (potentially due to a diseased enlarged heart being removed and replaced by smaller organ in a larger space), those with increased body surface area, previous cardiopulmonary bypass and increased duration of cardiopulmonary bypass.

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Case

A 58 year old man was referred with severe aortic stenosis with good left ventricular function causing dyspnoea NYHA II and angina CCS I. He underwent an uncomplicated metallic aortic valve replacement and was started on warfarin day 1 postoperatively.

His routine day 4 checks showed a small left sided pleural effusion and atrial fibrillation, which was then treated with amiodarone. Next morning whilst mobilising to the bathroom he became dizzy and the electrocardiogram (ECG) revealed sinus tachycardia. Chest x-ray and echocardiogram showed significant pericardial effusion which needed pericardiocentesis.

Post cardiac surgery pericardial effusion can be classified according to the time of presentation and aetiology as follows:

Immediate post operative	
Delayed	**Bleeding**
- days – weeks	*Bleeding*
	(anticoagulant use, pacing wire removal)
	Cardiac failure
	Chylopericardium
	Postpericardiotomy syndrome
- months	*Cardiac failure*
	Bleeding
	Postpericardiotomy syndrome

Aetiology

Immediate postoperative bleeding (Image 1 and 2)

Kuvin et al. (8), studied 4561 cardiac surgery patients retrospectively and found moderate to large pericardial effusions in 48 (1.05%); 36 of these developed cardiac tamponade. 8 of them were early i.e. within 24 hours of the operation.

Immediate post operative bleeding has been implicated in both early and delayed presentation of pericardial effusions (9). However, the focus here is on the immediate pericardial effusion.

The pericardium is often not closed intra-operatively and can communicate with one or both pleural spaces. Nevertheless, blood can localise to the pericardium causing effusions or compression of heart chambers impairing ventricular filling.

Patients who develop pericardial effusions and tamponade in the immediate post operative period often have early post operative bleeding (5) which is associated with higher chest drain outputs. Furthermore, on exploration of such patients most did not have a large single source of bleeding but rather had generalised capillary oozing from many small sites suggesting a generalised coagulopathy. Post operative bleeding therefore can be medical or surgical in origin.

Surgical bleeding pertains to the operative procedure i.e. from side branches of bypass grafts after CABG. Bleeding can also orginate from cannulation sites, aortic and paraaortic tissue, atriotomy sites, vent sites and from the bed of the internal mammary artery.

Medical factors relating to impaired coagulation include; the residual effect of heparin from an insufficient protamine sulphate dose after cardiopulmonary bypass; transfusion of heparinized pump blood, and thrombocytopenia. Cardiopulmonary bypass can also lead; to defective platelet function, haemodilution; increased plasminogen activation with subsequent fibronlysis and consumption of clotting factors all of which precipitate bleeding.

Pericardial effusions in the early post operative period causing high chest drain output are usually assumed to be secondary to bleeding. These have been associated with higher incidences in women and longer complex surgeries (8). The prolonged cardiopulmonary bypass time exacerbates the coagulopathy effect of bypass, similar to coagulopathy effects of bypass similar to a systemic inflammatory response syndrome (4).

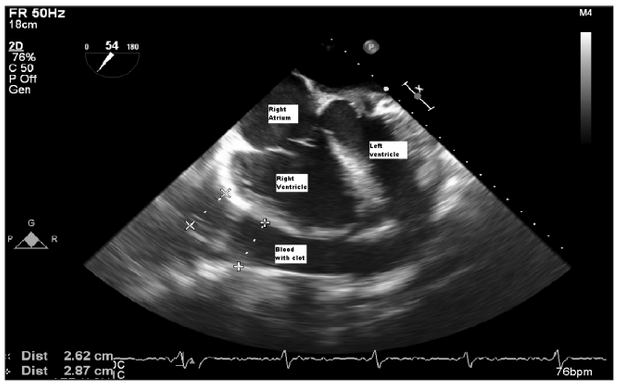

Image 1

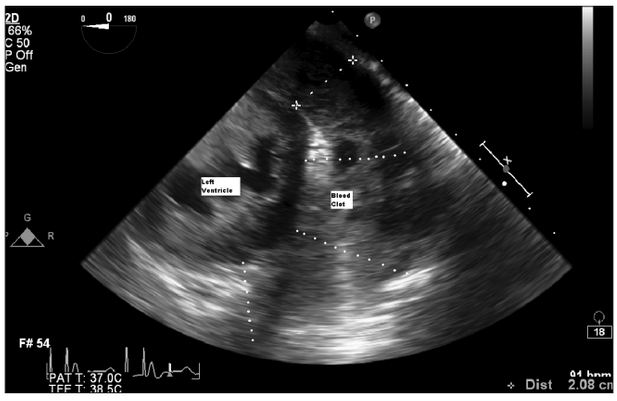

Image 2

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Anticoagulation/anti-platelet agents

The association between anticoagulation / antiplatelet use and pericardial effusions has been documented numerous times.

Pepi et al (1) studied 780 patients after cardiac surgery prospectively with echocardiography conducted prior to and 8 days post operatively. They found that significantly more patients undergoing CABG vs valve replacement had pericardial effusions and that small <5mm loculated or <9mm diffuse effusions were more common in patients after heart valve surgery, however moderate (<9mm located, <19mm diffuse) were found more often after CABG who are invariably on antiplatelet agents postoperatively. They also found that the incidence of cardiac tamponade in their patients post operatively was also significantly higher in those taking anticoagulants which was mostly in association with a valve replacement.

Malouf et al (10) investigated the role of anticoagulation in the development of pericardial effusions after cardiac surgery. They conducted a prospective study of 141 patients and found that there was a significant difference between the incidence of large pericardial effusions between those on anticoagulants and those on antiplatelet agents (aspirin, diypridamole). Within this group of patients, those that developed cardiac tamponade with a large pericardial effusion were all taking an anticoagulant. Monitoring patients anticoagulant levels they found that pericardial effusions were just as common in those within range as well as those beyond the therapeutic range, however larger effusions were statistically more common in those who were excessively anticoagulated (classed as INR >2.5), as well as the occurrence of tamponade in this group.

Tsang et al (5) studied 208 patients retrospectivey who had undergone 245 echo guided pericardiocentesis procedures for significant pericardial effusions or tamponade. Anticoagulation use was found to be a significant risk factor for the development of pericardial effusions less than 7 days postoperatively. Here, 86% of all 'early' effusions were attributed to this. Within these patients, over anti-coagulation was evident in the majority (INR range >3.5). Over anti-coagulation was also associated with 'late' (median 19 days, mean 39 days, range 8-212) pericardial effusions in 65% of these.

Pacing wires removal

The removal of temporary pacing wires has been associated with pericardial effusions and cardiac tamponade. Their use is commonplace after cardiac surgery for the pacing of atria and/or ventricles post-operatively and control of postoperative tachyarrythmias. In uncomplicated cases, these are usually removed on the fourth postoperative day.

Patients are then normally monitored for complications of pacing wire removal - among these are arrhythmias, injury to saphenous vein grafts; their side branches or graft clips, atrial and ventricular lacerations causing haemorrhage oreven cardiac tamponade. The degree of anticoagulation should always be considered prior to the procedure to reduce the risk of haemorrhage.

The incidence of pericardial effusion secondary to this is low. Omar et al. (2005) deduced the incidence of major complications after pacing wire removal to be 0.04%, with higher risks in redo cardiac surgeries or in anticoagulated patients. Mahon et al. (12) reviewed 22,717 patients after CABG and valvular surgery over 10 years at Cleveland Clinic, Ohio and showed an incidence of less that 1% of redo surgery for cardiac tamponade as a result of pacing wire removal. Interestingly, they found smokers to be at greater risk of reoperation after pacing wire removal.

Accepted practice is to apply gentle and sustained traction when removing wires, excessive and jerky force is more likely to cause complications. If there is significant resistance, the wires are cut at the skin surface and they retract internally.

Post pericardotomy syndrome (Image 3)

This is thought to be an autoimmune mediated inflammatory condition associated with pericardial and pleural effusions and rarely cardiac tamponade.

Underlying aetiology has been linked to both surgical trauma and autoimmune mediated reactions. In the case of pericardial effusions, the initial pericardial injury during surgery is thought to release pleuropericardial antigens and stimulate an immune response with inflammation and autoreactive reactions against the pericardium and pleura. This is similar to the process seen in in Dresslers syndrome post myocardial infarction.

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Ofori-Krakye et al. (6) studied 1290 patients post operatively and found postpericardiotomy syndrome in 21(1.6%) of these. Illness was defined as unexplained fever not related to infection or drug therapy, pericardial pain or friction rub and radiographic or echocardiographic evidence of pericardial fluid. Studies including Tsang et al. (5) have demonstrated postpericardiotomy syndrome as an important cause of late post operative pericardial effusions.

More recently, Imazio et al. (13) reported the incidence of postpericardiotomy syndrome to be 15%. They showed that pericardial effusions are prevalent in this population affecting 89% of these patients.

Onset of the illness ranged from days 7-186 (avg. 36 days) postoperatively, with symptom duration between 3- 372 days (mean of 41 days) (6). Five of these patients had a prolonged clinical course with frequent recurrences. Three developed late cardiac tamponade every time they presented with renewed pericardial inflammation. One required total pericardial stripping 372 days after the syndrome began to treat life threatening cardiac tamponade. All participants with 'late' (>7 days post op) onset cardiac tamponade had postpericardiotomy syndrome.

Borkon et al. (14) found that the concurrent use of anticoagulants alongside post pericardiotomy syndrome could predispose to haemorrhage and late pericardial effusions. There has also been some debate regarding the role of viral illnesses in the aetiology of the syndrome (15).

Chylopericardium

This is defined as a pericardial effusion consisting of chyle. This can be primary (of unknown cause) but more often secondary, owing to thoracic duct damage from surgery, malignancy (lymphoma), radiotherapy, subclavian vein thrombosis or infection (TB).

Diagnostic criteria to note include the milky opaque colour of the effusion. The triglyceride level is often greater than 500mg/dl and the ratio of triglyceride to cholesterol is <1. The pericardial fluid may also have a high protein, cholesterol,lactate dehydrogenase, fat globule and lymphocyte level.

Chylopericardium after cardiac surgery is rare, much of the literature focuses on individual case reports. Incidence is documented as 0.12% by Kan et al (16) and 0.22% by Campbell et al (17)

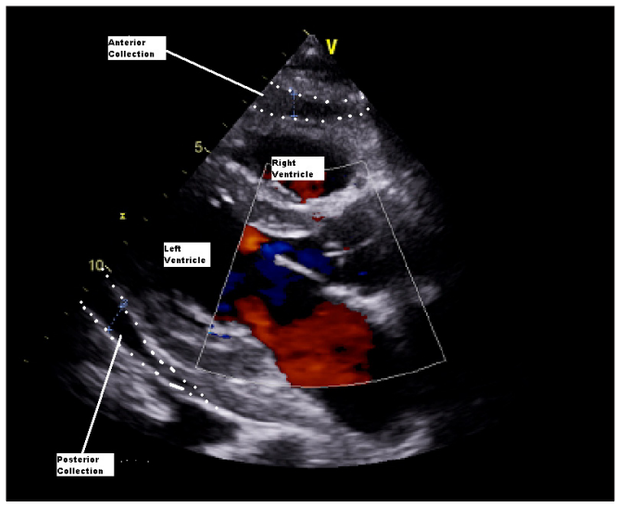


Image 3

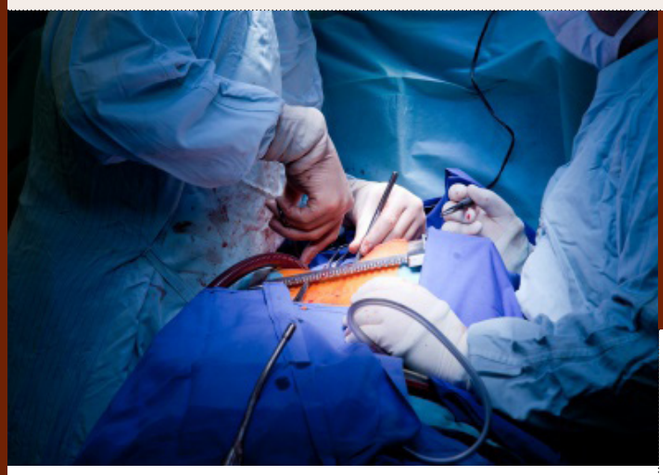
If the pleura is breached there is a possibility of direct injury to the main thoracic duct, but Cheng et al. (18) suggest damage to the thoracic duct from traction or when cross-clamping the aorta is unlikely, due to the anatomical location of the duct behind the pulmonary artery. Riquet et al (19) have suggested that inadvertent injury to a compromised right efferent lymphatic duct within the pericardium can cause chyle backflow leading to chylopericardium. Kan et al (16) mention that high dissection of the internal thoracic artery requiring pleural dissection also risks damage to the anterior mediastinal node chain which can result in chylopericardium. In children chyle leak from the thymus has also been suggested as a cause postoperatively.

Clinical presentations (signs and symptoms)

Not all pericardial effusions present as soon as they develop. Presentation has been described as acute (5, 9) or insidious in onset. The visceral pericardium's mesothelial cells produce pericardial fluid. There is normally 15-50mls present at any one time. This fluid is actively cycled by lymphatic drainage. Symptoms occur as the pericardial pressure is affected by the volume and rate of fluid accumulation and pericardial membrane compliance. If there is a slow and gradual accumulation of fluid, a compensatory stretching of the pericardium and maintenance of adequate lymphatic allows pericardial pressures to remain the same, or increase slightly and gradually. In contrast, a sudden increase in fluid volumes does not allow these compensatory measures, leading to a more acute presentation.

PERICARDIAL EFFUSIONS & COLLECTIONS AFTER CARDIAC SURGERY

E Lizen, AV Singh



Acute:

- *Most often with bleeding*
- *Specific sign of raised drain output >200mls/hour*
- *Sudden cessation of bleeding due to blocked drains is more often associated with cardiac tamponade (9)*
- *Tachycardia*
- *Hypotension*
- *Low cardiac output (cold peripheries, decreased urine output, metabolic acidosis)*

Insidious:

- *Malaise*
- *Dyspnoea*
- *Fever*
- *Chest pain*
- *Oedema feet*
- *Presyncope, syncope*
- *Nausea, vomiting, abdominal pain*

Clinical diagnosis of postpericardiotomy syndrome relies on the presence ≥2 of the following findings to fulfill recent diagnostic criteria (20):

- *Fever lasting beyond the first postoperative week with no infection (local or systemic)*
- *Pleuritic chest pain*
- *Friction rub*
- *Pleural effusion*
- *Evidence of new or worsening pericardial effusion*
- *Presence of ≥2 of these findings full fill recent criteria for diagnosis.*

Pericardial effusions & collections after cardiac surgery

Cardiothoracic & Critical Care

Large pericardial effusions can cause compression of the base of the left lung; crackles and reduced breath sounds may manifest as a result of this. Regardless of pericardial effusion size, a patients pulse, JVP, and BP are maintained unless the intrapericardial pressures jeopardise ventricular filling. Deterioration of these parameters increases the risk of cardiac tamponade.

Investigations

Diagnostic - Echocardiogram – transthoracic or transoesophageal

Signs of haemodynamic compromise include right ventricular (anterior wall), right atrial or rarely left atrial diastolic collapse and plethora/dilatation of the IVC without appropriate respiratory variation.

Transoesophageal echocardiogram provides better views early post operatively compared to transthoracic echocardiograms. This is especially useful for loculated pericardial effusions and when there are restricted transthoracic windows due to; overlying bandages, obesity, obstructive lung disease, mechanical ventilation, limited patient positioning, and incision site tenderness.

Other - Chest x-ray

Enlarged cardiac sillehoute typically globular in insidious cases, concomitant pleural collection.

Electrocardiogram

Sinus tachycardia, smaller QRS complex, electrical alternans showing cyclical QRS amplitude variation associated with large effusions or tamponade.

Computed Tomography and Magnetic resonance imaging

Have value in insidious situations to assess for additional pathology like cardiac tumours and assessment of myocardium.

Pericardial fluid

From pericardiocentesis for chylomicrons, gram stain and culture will clinch diagnosis in patients with chylous effusion and purulent collections.

PERICARDIAL EFFUSIONS & COLLECTIONS AFTER CARDIAC SURGERY

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Colour of the fluid gives a clue:

• Clear/serous fluid - transudate, low protein level and a low cell count – cardiac failure.
• Serosanguinous - bleeding, postpericardiotomy syndrome, neoplasm.
• Purulent - infection - mediastinitis.
• Blood and non-clotting – associated with anticoagulants.

Bloods

• Full blood count - low haemoglobin.
• Raised white cell count and polymorphonuclear cells may also suggesting post-pericardiotomy syndrome or infection.
• Clotting – anticoagulant effect suggesting cause and in preparation for intervention.
• Elevated ESR - post pericardiotomy syndrome.

Management

Depends largely on the patients clinical status.

Unstable patient post-operatively

Severe hypotension or cardiac arrest warrant emergency re-sternotomy.

Stable patient

Medical

- Reversal of anticoagulation by Vit K, plasma or octaplex (prothrombin complex concentrate)
- Optimisation of antifailure treatment
- Anti-inflammatory drugs for postpericardiotomy syndrome
- Low fat diet, oral octreotide and pareneteral nutrition for chylopericardium

Percutaneous Intervention

Echocardiography guided pericardiocentesis for drainage and leaving a pigtail catheter in for further aspiration is most appropriate for non-loculated and suitably located effusions.

Surgical

Re-sternotomy

This is more often used in patients who have evidence of cardiac tamponade, haematoma or ongoing evidence of bleeding the early postoperative period. Re-sternotomy may also be considered in patients with loculated posterior effusions which are difficult to access percutaneously.

Thoracic duct ligation

This may have to be performed rarely via thoracotomy or video assisted thoracoscopic surgery for failure of conservative treatment of chylous pericardial effusions.

Questions

1. With respect to pericardial collection in the immediate post-operative period, which of the following is true:

(A) Is most often due to a surgical bleed

(B) Will always manifest with increased drain output

(C) Chest x-ray is the best investigation for diagnosis

(D) Prolonged bypass time increases the risk of pericardial collections.

2) Which of the following is false?

(A) The risk of pericardial effusions after pacing wire removal is small, but patients should be monitored carefully during and after removal.

(B) The most common ECG finding in pericardial effusion is sinus tachycardia

(C) There is a direct correlation between the incidence of pericardial effusion and the degree of over anti-coagulation in patients.

(D) It is reasonable to treat small, haemodynamically stable pericardial effusions conservatively.

3) At 8 days post operatively after CABG Mr Smith develops a fever and feels a little tired. Cultures are negative and he appears a little more breathless than usual on exertion. The CXR is normal and echocardiography has shown a 1cm posterior pericardial effusion. Which of the following aetiologies is most likely?

(A) Bleeding secondary to use of aspirin

(B) Bleeding from pacing wire removal

(C) Post-pericardiotomy syndrome

(D) Heart failure

4) Which of the following investigations is the most appropriate in an intubated patient in the immediate post-operative period who has a suspected pericardial effusion?

(A) CXR

(B) ECG

(C) ECHO

(D) TOE

PERICARDIAL EFFUSIONS & COLLECTIONS AFTER CARDIAC SURGERY

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Pericardial effusions & collections after cardiac surgery
Cardiothoracic & Critical Care

5) A 70 year old gentleman, day 4 post-op mechanical aortic valve replacement develops shortness of breath, malaise and tachycardia and cold peripheries. His VBG shows a lactate of 4, his bloods show a Hb of 88, ECHO showed a 3cm non-loculated circumferential pericardial collection with impaired right atrial and ventricular filling. Management should be...

(A) Immediate resternotomy

(B) Urgent pericardiocentesis

(C) Reverse warfarin and proceed to pericardiocentesis

(D) Conservative management

Answers

1. D

Surgical factors eg, bleeding from side branches of a graft do play a role however, more often the specific cause for bleeding is not located. Instead there appears to be a impaired state of coagulation and general oozing from multiple sources. Increased drain output is a presenting feature of immediate postoperative pericardial effusions however sometimes drains may be blocked and this sign might not be evident. CXR's are more likely to show changes in association with chronic pericardial effusions. Prolonged bypass time is associated with increased risk of immediate postoperative bleeding and pericardial effusions.

2. C

There was no evidence to suggest over anti-coagulated patients were more likely to have pericardial effusions compared to those within the therapeutic range. There is a suggestion that larger pericardial effusions present in patients whose INR is beyond the therapeutic range.

3. C

Post pericardiotomy syndrome- This is quite prevalent in the postoperative period. The significance of the pericardial effusion varies. There is often fever, together with tachycardia, dyspnoea and a pericardial friction rub. It is managed depending on the stability of the patient; drainage if tamponade is present, and anti-inflammatory use is commonly used as medical management of this condition.

4. D

TOE - Postoperatively, transthoracic echocardiograms are limited for numerous reasons; pericardial effusions are often loculated and there are poor window views to assess this transthoracically. As that patient is intubated TOE is most appropriate. A CXR is a useful initial investigation but gives little detail at this stage.

5. C

Reverse warfarin and proceed to pericardiocentesis - this is the most appropriate in this case as the effusion is causing cardiac tamponade. It is large enough for pericardiocentesis. And as it is non-loculated this procedure is more likely to be successful. It is still an invasive procedure which requires normal coagulation. Resternotomy should be contemplated in situations where pericardiocentesis is more likely to fail i.e. posterior or loculated collections.

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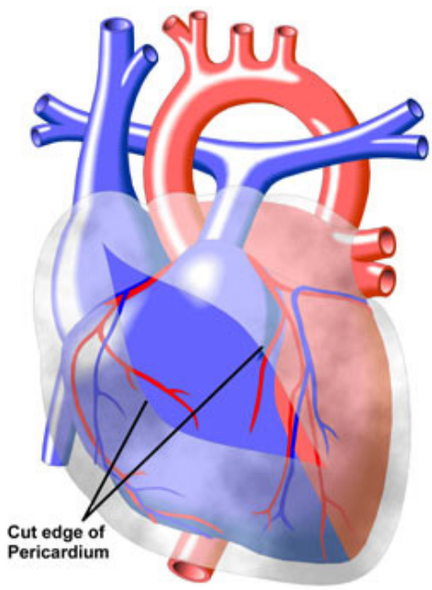

PERICARDIAL EFFUSIONS & COLLECTIONS AFTER CARDIAC SURGERY

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REVIEW OF PRIAPISM FOR CORE SURGICAL TRAINEES

TL Lewis, N Bedi, A Deshpande

Review Of Priapism For Core Surgical Trainees Urology

Abstract

Priapism is an involuntary penile erection that lasts for longer than four hours, unrelated to sexual stimulation. It is a rare condition that requires urgent surgical treatment to prevent permanent impotence. This article highlights common causes of priapism and introduces readers to the anatomy and physiology of penile erection. Priapism can be categorised into 'High flow', 'Low flow' or rarely 'Stuttering'. Diagnosis and emergent management of these different types of priapism are discussed. Options to create a surgical shunt to manage refractive cases are reviewed alongside potential complications.

Keywords: *Priapism, Urology Emergency, erectile dysfunction, penis anatomy.*

Introduction

Priapism is an involuntary penile erection that lasts for longer than four hours, unrelated to sexual stimulation. It is a rare condition, that requires urgent surgical treatment to prevent permanent impotence (1).

Causes

Priapism was first reported by Callaway in 1824 (2–5), and current data shows that it is relatively uncommon, with an incidence of 1.5 per 100,000 (6,7).

CAUSES OF PRIAPISM	
Idiopathic	
Vasoactive drugs	Drugs for intracavernous injection Anticoagulants Antihypertensives CNS-acting drugs
Haematologic disorders	Sickle Cell disease Thalassemia Thrombocythemia Multiple myeloma Leukaemia Paroxysmal nocturnal haemoglobinuria Anaemia
Metabolic disorders	Renal Failure Amyloidosis Fabry's Disease Gout Diabetes Nephrotic Syndrome
Trauma	Usually to the perineum
Tumours	Especially pelvic malignancy
Neurological disorders	
Inflammation	
Kawasaki Disease	

Causes Of Priapism Table

Physiology Of Erection

Penile erection is an interplay between physical & psychological processes and results from cavernous, arterial & arteriolar smooth muscle relaxation which increases arterial inflow and restricts venous outflow (5,8).

Sexual stimulation causes neurotransmitter release from cavernous nerve terminals which dilates arterioles & arteries resulting in increased blood flow to the sinusoids. Relaxation of the trabecular smooth muscle dilates the sinusoids and enables expansion of the erectile tissue. Venous outflow is reduced by compression of subtunical venules as a result of the expansion of the erectile tissue against the tunica which encloses the emissary veins between its two layers.

Anatomy

The main anatomical components of priapism are the arterial system, the venous system and the corpora cavernosa as highlighted above.

Arterial supply

The predominant source of blood supply to the penis is the internal pudendal artery, a branch of the internal iliac artery. However, accessory internal pudendal artery may also arise from the external iliac, femoral, obturator or vesical arteries and can be found in conjunction with internal pudendal artery in 75% of cases. The internal pudendal artery continues as the common penile artery after giving off a branch to the perineum.

The common penile artery divides into the dorsal artery of the penis & the bulbourethral artery. The dorsal artery gives off the cavernous artery which travels through the centre of the corpus cavernosum on each side & gives off helicine arteries. The dorsal artery continues on the dorsal aspect of the corpora cavernosa giving off circumflex arteries & ends up supplying the glans penis. The bulbourethral artery supplies the corpus spongiosum & its bulb as well as the urethra & anastomoses with the dorsal artery at the coronal sulcus.

Venous Drainage

The venous drainage of the penis commences from the corporal sinusoids as tiny venules just beneath the tunica albuginea. These venules form the subtunical venous plexus in the trabeculae beneath the tunica albuginea & exit through it as the emissary veins.

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The emissary veins from the corpora drain dorsally into the deep dorsal vein, laterally to the circumflex veins & ventrally into the periurethral veins. The deep dorsal vein is the main venous drainage of the penis and eventually drains into the periprostatic venous plexus. The proximal corpora cavernosa drain via the emissary veins into the cavernosal & crural veins, which join the periurethral veins & drain into the internal pudendal veins. The skin of the penis is drained via the superficial dorsal veins into the saphenous veins.

Pathophysiology And Classification

Priapism results from failure of detumescence, which is directly related to decreased venous outflow or increased arterial inflow.

Priapism can be classified as either 1) low flow or ischaemic, 2) high-flow or arterial (9) or 3) 'stuttering' (rare). Low-flow priapism is characterised by a painful hard erection with a corresponding soft glans and corpus spongiosum(rare exception). This is caused by occlusion of the venous outflow which in turn leads to blood-pooling, clotting and resulting ischemia. Sustained ischaemia can lead to tissue fibrosis and permanent loss of erectile function. Conversely, high flow priapism results from cavernous arterial inflow, which is typically characterized by sustained painless erection.

Diagnosis

Diagnosis of priapism is made clinically by history and physical examination. Intracorporeal blood gas analysis is crucial to ascertain whether this is low-flow or arterial as a low-flow priapism is a true urological emergency (7,10,11).

Typical Blood Gas Values

Source	PO2 (mm Hg)	PCO2 (mm Hg)	pH
Ischemic priapism (cavernous blood)[3]	<30	>60	<7.25
Normal arterial blood (room air)	>90	<40	7.40
Normal mixed venous blood (room air)	40	50	7.35

Typical Blood Gas Values Table (10)

Management

Indications For Treatment

Ischaemic priapism, if left untreated for more than 4 hours, will commonly result in permanent impotence (1,12). As a result it should be considered a urological emergency and treatment initiated as soon as possible.

Investigations

The single most important investigation to carry out is intracorporeal blood gas analysis to distinguish between the two types as described above. Other investigations are rarely indicated in clinical practice. These include Doppler study and duplex sonogram of the penis to determine high flow priapism. Cavernosography is no longer used due to subsequent risk of damage to the erectile tissue.

Treatment

Treatment for priapism depends on the type of priapism. It is important that a penile block (usually 5-10ml of 0.5% Marcaine) be administered right at the start to provide patient with adequate analgesia.

Low-Flow Priapism

If the patient presents within 4 hours, then physical methods e.g. asking patient to ejaculate, cold showers, walking up and down the stairs, etc can be employed. Oral terbutaline, a Ð2 agonist, may help in intracavernosal injection-related cases, administered in 5 – 10 mg dose (14). Failure of these methods requires intracorporeal aspiration (up to 100ml) with 19 – 21 gauge butterfly needle, inserted through the side shaft of each cavernosum. The success rates of aspiration alone vary between 24 – 36%.

For recurrence or failure after aspiration, the patient needs to be shifted to a monitored bed as intracavernosal Ð-adrenergic agonist is required. The drug with the least potential cardiovascular side-effects is phenylephrine; 200 mcg (diluted down to 1 ml) is administered every 5 min until detumescence or 1 mg total is reached. Other drugs which can be used include ephedrine, epinephrine, norepinepherine and metaraminol. Also at this point, the nearest specialist centre should be contacted, as the patient may need referral for shunt surgery if there is no response.

In the specialist unit, a biopsy & shunt surgery is performed proceeding from distal shunts to proximal shunts with reported success rates, but with erectile dysfunction in up to 90% of cases (10).

The last step in the management of priapism after failure of all of the above steps is insertion of penile prostheses, hence the importance of corporal biopsy at the same time as shunt surgery, to assess tissue necrosis or fibrosis. If corporal fibrosis is noted on biopsy, penile prostheses can be inserted to prevent penile shortening.

Treatment of priapism secondary to sickle cell anaemia follows an additional pathway and relies on hydration, analgesia and blood exchange transfusion. Intravenous fluid therapy aims to inhibit sickling by decreasing tonicity and improving circulation. Regular blood transfusions should take place until haemoglobin levels are >10 mg/dl and haemoglobin S < 30%(1).

High-flow Priapism

This is not an emergency & can be managed expectantly. The most common cause for high-flow priapism is the formation of an arterio-venous fistula. Compression can be applied over the fistula after identification with Doppler ultrasound.

For those patients with ongoing symptoms, diagnosis and treatment can be offered by angiography +/- embolization, using absorbable materials e.g. autologous clots or absorbable gels. There is about 5% risk of erectile dysfunction with the absorbable materials as compared to nearly 40% with non-absorbable materials (10).

In case of a definite fistula being demonstrated, open exploration & repair of the fistula can be attempted.

REVIEW OF PRIAPISM FOR CORE SURGICAL TRAINEES

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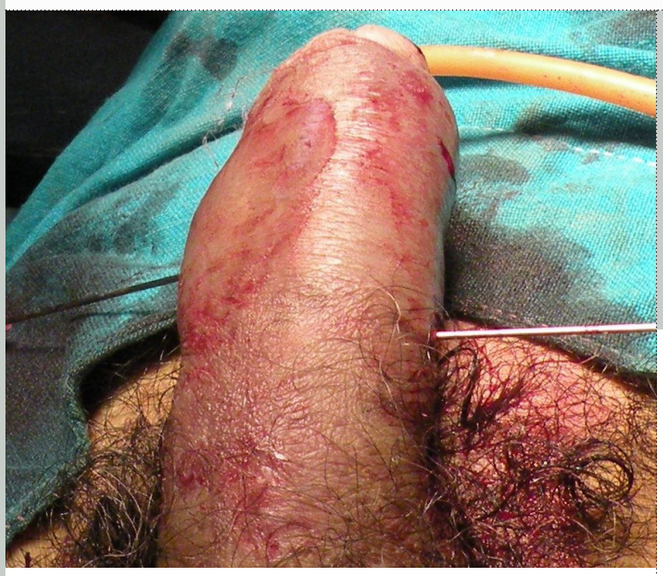

Shunt Surgery

As mentioned above, in case of failure of intra-cavernosal injection of an Ð-adrenergic agonist, shunt surgery is performed & there are various techniques described in the literature (1,8,12,15–21). In case of low-flow priapism, the primary objective is to create a new venous outflow and thereby help secure normal arterial activity in the corpora cavernosa.

Shunts can be broadly classified as distal & proximal. Distal shunts rely on shunting blood from the affected corpora cavernosa into the uninvolved corpus spongiosum. Winter's shunt involves inserting a Tru-cut needle through glans into the corpus cavernosum while Ebbehoj shunt is achieved by insertion of a scalpel. There is a variation of the scalpel technique called the Lue shunt which involves a 900 rotation of the scalpel resulting in a T-shunt. El-Ghorab shunt involves excision of an ellipse of the tunica albuginea on each side at the tip of the corpus cavernosum with a dorsal transverse incision distal to the coronal ridge (10, 18-20).

When distal shunts fail, proximal shunts should be considered, usually in a specialist unit setting with the appropriate expertise & experience. Quackels described a proximal cavernoso-spongiosal shunt where a 1 cm long ellipse of each structure is excised & they are then anastomosed together, with care taken to avoid urethral injury. Grayhack shunt involves a cavernoso-saphenous anastomosis where the long saphenous vein is mobilised & sutured to the corpus cavernosum. This technique is more difficult but reduces the risk of urethral injury (10,17,21).

Review Of Priapism For Core Surgical Trainees Urology

Following surgery, it is important to establish a number of key physiological and physical parameters to reduce the risk of recurrent priapism. These are:

- Intracorporeal blood pressure must remain lower than 40mmHg for 10 min.
- Intracorporeal blood gases should normalise.
- Avoid circular compression dressings as these can obstruct venous outflow.

An ischaemic blood gas post-operatively indicates recurrent priapism with a failed shunt and therefore it is appropriate to consider further surgical intervention.

Complications

There are a range of complications of surgical cavernoso-spongiosal shunts including cavernous-glanular fistula, which can cause veno-occlusive dysfunction and possible impotence. Other surgical complications include: stricture formation, urethro-cavernous fistula, urethro-cutaneous fistula, gangrene, necrosis, abscesses, cellulitis and fibrosis leading to a shortened penis.

Conclusion

Priapism is a urological emergency, which can be broadly classified as either low-flow or high-flow. Diagnosis is usually clinical although intracorporal blood gas analysis is vital to assess the severity and classification of the priapism. There are other investigations if required although these are typically not used in clinical practice. The primary treatment for priapism is medical therapy with intracavernosal washout ± Ð-adrenergic agonists. There are surgical shunt procedures if the priapism does not respond to medical therapy but are best managed in specialist centres with adequate expertise & experience. Penile prostheses form the last line of management if corporal biopsies show fibrosis.

Case Vignette

Mr Smith is a 60 year old warehouse worker who comes to A&E with an erection persisting 6 hours prior to his arrival. He complains of pain and passing blood in the urine, and reveals that urinating is difficult and painful. There are no other associated symptoms. His last normal erection was three days before this began. He denies taking any erectile enhancing substances, and does not have sickle cell anaemia. This has never happened before. There is no reported injury to the genital region.

REVIEW OF PRIAPISM FOR CORE SURGICAL TRAINEES

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How are you going to manage this patient as the on call SHO?

Answer:

- Inform the on-call Urology registrar. (Urological emergency)
- Give oral vasodilators.
- Aspirate intracorporeal blood sample and analyse blood gases to classify type of priapism.

MCQs

1. What is the most common cause of paediatric priapism?

A. Idiopathic
B. Sickle cell disease
C. Trauma
D. Kawasaki's Disease

2. If the priapism fails to respond to medical therapy what is the initial surgical shunt option?

A. Winter's procedure
B. Quackels procedure
C. Grayhack's procedure

3. Which one of the following is the rarest complication of surgical shunting?

A. cavernous-glandular fistula
B. stricture formation
C. urethra-cavernous fistula
D. urethra-cutaneous fistula
E. Pulmonary embolism

Answers

1. B
2. A
3. C

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JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

C Saxby, R Williams, H Khalil

Juvenile Nasopharyngeal Angiofibroma
Neurosurgery

Abstract

Juvenile Nasopharyngeal Angiofibroma (JNA) was first described by Hippocrates in the 5th century BC (1). In 1906 Chaveau introduced the term "juvenile nasopharyngeal fibroma" (2). After histology studies on operative specimens in 1940, Friedberg changed the name to "angiofibroma" (3).

JNA originates in the posterior nasal cavity, near the back of the middle turbinate (4). It is a rare and locally invasive vascular tumour. JNA is histologically benign; however it may mimic malignancy by its progressive growth and destruction of surrounding tissues. This can lead to a significant degree of morbidity, related to intracranial extension or massive haemorrhage (4).

Keywords: *Juvenile nasopharyngeal angiofibroma; paediatric rhinology.*

Aetiology

JNA account for less than 0.5% of all head and neck tumours (5). The tumour is almost exclusively found in male adolescents, however cases have been reported in males aged over 25 years and females (6). The mean age range is between 14 to 17 years (7). JNA appear to be more common in the Middle East and Indian subcontinent (5).

Pathophysiology

JNA are benign fibro-vascular lesions. Macroscopically they appear sessile and lobulated and red-pink to tan in colour (shown in figure 1) (8). JNA are composed of a proliferating and irregular vascular tissue within a fibrous stroma containing collagen and fibroblasts and are surrounded by a fibrous pseudo-capsule (9). JNA contain multiple vascular channels which are thin walled and lack smooth muscle and elastic fibres, thereby leading to the possibility of life threatening epistaxis (10).

The diagnosis of JNA is based on the site of origin. JNA arise on the posterolateral nasal wall at the sphenopalatine foramen (10). The internal maxillary artery is the most common vascular source from which JNA arise (11).

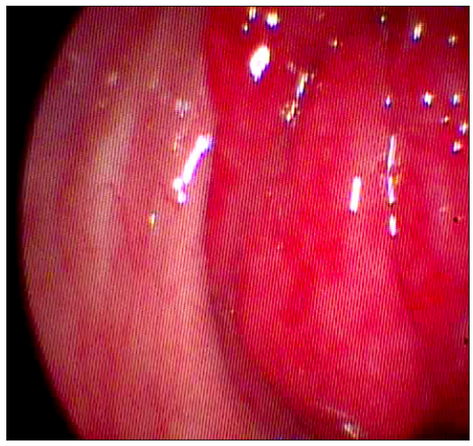

Figure 1: Endoscopic view of a JNA.

A hormonal influence of JNA has been speculated since it is a tumour almost exclusively of adolescent males. Montag et al in 2006 noted androgen and oestrogen receptors on some JNA cells (12). There has, however, been no endocrine abnormality identified among individuals with JNA (13, 14).

A recent hypothesis by Schik states that JNA are a vascular malformation resulting from incomplete regression of the first branchial arch artery which connects the internal and external carotid arteries during embryonic development (15). Other theories suggest a role for vascular endothelial growth factor receptor-2, transforming growth factor beta-1, or insulin-like growth factor- 2 (16).

Clinical features

Commonly JNA present with progressive unilateral nasal obstruction and recurrent epistaxis in an adolescent male (10). On examination a lobulated, smooth pink nasal mass may be seen. Headache and facial pain may arise secondarily to the blockage of paranasal sinuses. Unilateral otitis media with effusion may occur due to the impairment of Eustachian tube function (5). With more advanced tumours proptosis, cheek swelling and altered vision may occur, indicating an involvement of the orbit. Intracranial extension of JNA can lead to headaches and cranial neuropathies (10).

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Imaging

Imaging is very important and is often used to make the diagnosis of a JNA since biopsy carries the risk of massive haemorrhage (9). To assess pre-operative extension both computed tomography (CT) and magnetic resonance imaging (MRI) are required; the CT scan to show bone changes and the MRI to look at soft tissue extension (9). Advances in radiological imaging have significantly contributed to better preoperative management and treatment planning.

Key features to support a diagnosis of JNA include the presence of a vascular mass at the posterior nasal cavity (shown in figure 2). Enlargement of the sphenopalatine foramen with erosion of the pterygoid plates are regarded as pathognomonic radiological features for JNA (9).

The high rate of recurrence makes post-operative surveillance an important part. The presence and extent of invasion of the sphenoid is very important since it is the main determinant of recurrence (17).

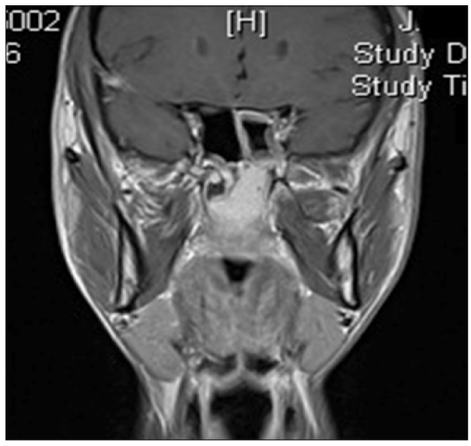

Figure 2: Coronal CT scan showing a JNA in the posterior nasal cavity.

Staging

Staging systems were developed mainly to predict recurrence and to compare surgical results. They are based upon the behaviour of the tumour, the routes of spread, difficult areas for surgical exposure, and possible sites of recurrence (8).

Preoperative tumour stage is very important in predicting the prognosis of JNA patients. There are many described staging systems including Fisch's and Chandler's; however the Radowski classification is the most widely used (8).

The Radowski Staging System for JNA	
Stage I	Ia - limited to nasal cavity/ nasopharynx Ib - involvement of at least 1 paranasal sinus
Stage II	IIa - minimal extension into pterygomaxillary fossa IIb – fills pterygomaxillary fossa +/- extension into orbit IIc - extends into infratemporal fossa
Stage III	IIIa- skull base erosion with minimal intracranial extension IIIb – skull base erosion with extensive intracranial extension +/- cavernous sinus

Treatment

Surgery is considered the treatment of choice for JNA (18). In the last two decades there has been a shift towards endoscopic techniques. Ardehali et al recommended endoscopic resection of JNA to be the first surgical step for tumours with stages I to IIIa (19). This was due to the significantly lower intraoperative blood loss, shorter operative time and recurrence rates in comparison to traditional approaches such as midfacial degloving (20). However endoscopic resection has been reported to limit tumour mobilisation within the nasal cavity and with brisk haemorrhage can lead to a significant impairment of vision (20, 21).

Since JNA are highly vascular tumours bleeding during surgery is a critical subject. Studies have compared the blood loss between endoscopic and external approaches, showing a lower loss in endoscopic surgery (20, 21). Ardehali et al reported a decrease in intraoperative and postoperative haemorrhage when surgical resection was combined with preoperative embolisation (19). Pre-operative embolisation is routinely used to minimise blood loss, which can be catastrophic otherwise. However, preoperative embolisation can obscure tumour borders and therefore complicate resections (22).

The use of radiotherapy in JNA is often reserved for advanced unresectable tumours and failure of complete tumour removal. McAfee et al treated 22 patients affected by high staged JNA with radiotherapy (10 cases as primary treatment, and in 12 for recurrence).

Local control was obtained in 90% of patients (23). Late complications, which occurred in 32% of cases, included cataracts, transient central nervous system syndrome, and cutaneous basal cell carcinoma (23).

JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

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Several studies on hormone pathogenesis have demonstrated the hormonal dependence of this tumour, suggesting a promising role of oestrogen or androgen receptor blockers in its treatment (24, 25). Gates et al administered flutamide, a non-steroidal androgen receptor blocker, in 5 patients with a diagnosis of JNA and detected an average tumour regression of 44% in four cases (24). However, Labra et al observed no significant differences between tumour size before or after flutamide administration in a report of 7 patients (25).

Recurrences can occur as early as three to four months following surgery and recurrence rates of 39.5% have been reported (17). Therefore extensive surgical resections and long term follow up of patients is required.

Conclusion

JNA is a locally aggressive and histologically benign vascular tumour primarily affecting adolescent males. Various treatment modalities have been discussed in the management of JNA. Surgery constitutes the primary treatment option however there are no standard guidelines available in the literature. Treatment depends on doctor and patient preferences and the available expertise. As JNA is a rare tumour it is important to evaluate for other neoplasms that may also manifest in the nasal cavity.

MCQs

Please state if the following are True or False:

1. JNA:

a) commonly arise from the maxillary sinus
b) can extend intracranially
c) arise on the posterolateral nasal wall
d) are histologically malignant tumours

2. The clinical presentation of JNA may include:

a) recurrent epistaxis
b) night sweats
c) proptosis
d) unilateral nasal obstruction

3. JNA:

a) account for around 5% of all head and neck tumours
b) require an urgent biopsy to confirm diagnosis
c) both a CT scan and MRI scan are required before surgery
d) almost exclusively found in female adolescents

4. In the treatment of JNA:

a) pre-operative embolisation is rarely utilised
b) surgical resection is only recommended for tumours limited to the nasal cavity
c) radiotherapy is often reserved for advanced unresectable tumours
d) long term follow up is required due to high recurrence rates

5. JNA may be staged using the following system:

a) Radowski
b) Chandler
c) House Brackmann
d) Jackler

Answers

1. FTTF
2. TFTT
3. FFTF
4. FFTT
5. TTFF

JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

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BIG TUMOUR, BIG SUCCESS, WILMS' TUMOUR – A REVIEW

A Jones, S Wood, R Craigie

Big Tumour, Big Success, Wilms' Tumour – A Review
Paediatric Surgery

Abstract

Wilms' Tumour or Nephroblastoma is the fifth most common malignancy in children, and accounts for approximately 5% of all childhood cancers. In this review we give an outline of the pathophysiology, investigation and treatment of paediatric Wilms' tumour for the core surgical trainee. Case studies, imaging and multiple choice questions allow the reader to engage and digest the important points.

Keywords: *Wilms' tumour, nephroblastoma, paediatric tumour.*

Introduction

Nephroblastoma, more often known by its eponym Wilms' Tumour, is the most common renal tumour of childhood and accounts for approximately 5% of all childhood cancers. Although the first recorded nephrectomy for a renal tumour in a child was in 1877 by Dr Thomas Jessop at Leeds General Infirmary [1] it was not until 1899 that Max Wilms, Professor of Surgery in Heidelberg, described the tumour that now bears his name.

Wilms tumour is a wonderful example of the significant improvements in cancer therapy that we have seen in the last half a century. In the 1930's, long term survival rates were approximately 10%, but with the introduction and refinement of chemotherapy and radiotherapy, these have improved dramatically to more than 90%. More recent changes to therapy have been concerned with reducing toxicity and long term effects of treatment, as well as improving survival. Surgery remains a key part in the management of children with Wilms' tumour.

There are around 100 new cases of Wilms' tumour in the UK each year, the majority of which are in children under 5 years of age. It has been known in older children and adults, but then, it is more likely to occur in the presence of an underlying syndrome or genetic abnormality.

Genetics and tumour biology

The syndromic associations of Wilms' tumours (table 1) have enabled investigators to discover genetic loci which both predispose to the tumour formation, and influence prognosis. The discovery of a constitutional deletion in 11p13 in children with WAGR syndrome[2] led to consideration of this change being the first 'hit' in the Knudson and Strong "two hit" genetic model. Tumour formation consequent to loss of the second copy of this gene led to the suggestion that it was a tumour suppressor gene, named WT1. WT1 has an important role in the control and differentiation of various organs, and has been shown to regulate the expression of other genes, including growth-inducing genes such as insulin-like growth factor-2 (IGF-2). WT1 is known to bind to, and stabilise the tumour suppressor gene p53, possibly resulting in the increase in wild type p53, often seen in Wilms' tumours[3].

A second locus, WT2, has been identified on chromosome 11p15.5, which when constitutional, causes Beckwith-Wiedemann syndrome. WT2 is associated with bilateral and familial Wilms' tumours.

The WT1 gene is identified in only 5-10% of non syndromic Wilms' tumours and, along with WT2 appears to have no prognostic significance. Denys-Drash syndrome is associated with constitutional abnormalities of the WT1 gene with 50-90% of children developing Wilms' tumours.

Familial cases of Wilms' tumours account for approximately 1-5%. Analysis of 7 different kindreds demonstrated links with chromosome bands, 17q12-21, and 19q13.3-q13.4[5,6] as well as WT2.

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In tumours where WT1 and WT2 were normal (2 alleles present), further analysis revealed additional Wilms' loci on chromosomes 16q and 1p. Grundy analysed 232 tumours and found loss of heterozygosity (LOH) at 16q in 17% and 1p in 12%. Data from the NWTS-4 trial suggested that LOH in either of these chromosomes predicted an adverse outcome. This was proved in NWTS-5 where combined LOH in 1p and 16q was an adverse indicator in all stages of Wilms' tumour. Increased expression of the p53 protein is also associated with advanced stages of presentation, which have a poorer prognosis. These biogenetic risk factors continue to be studied in current trials, in the context of treatment toxicity and the limitation of adjuvant therapies.

Associated Syndrome	Features
Beckwith-Wiedemann Syndrome	Organomegaly, Macroglossia, ear creases, hypoglycaemia, abdominal wall defects, seizures, cryptorchidism, increased risk of many solid tumours.
WAGR Syndrome	**W**ilms tumour, **A**niridia, **G**enitourinary anomalies, mental **R**etardation
Denys-Drash Syndrome	Gonadal dysgenesis, Wilms tumour, Nephropathy

Table 1: Syndromes associated with Wilms' tumour formation.

Wilms tumours are classically 'triphasic' on histological examination. They possess three cell types - blastemal, stromal and epithelial. They may have an element of necrosis (generally a good prognostic factor), and as with most malignancies, the extent of cell differentiation is important to note. The more anaplastic, the higher risk the tumour is deemed.

Nephrogenic rests (NR) are foci of embryonal cells which abnormally persist after thirty six weeks of gestation. They either regress to fibrous tissue, or progress to nephroblastoma. NR are found in 1% of perinatal post mortem kidneys, but in 25-40% of patients with Wilms' tumours[7]. They are therefore considered precursors of Wilms' tumour. There are two major distinct categories, perilobar, associated with Beckwith-Wiedemann syndrome, and intralobar, associated with WAGR and Denys-Drash. Hyperplastic rests should be treated as Wilms' tumour.

Clinical Features

Typically, children with Wilms' tumours present at a mean of 3 years of age with a large asymptomatic abdominal mass. Other clinical features may include haematuria, abdominal pain, fever, hypertension and varicocoele. Rarely, the child can present shocked and peritonitic as a result of a bleed within the tumour, or tumour rupture. Potential differential diagnoses are listed in table 2.

Differential Diagnosis	
Benign	**Malignant**
Xanthogranulomatous pyelonephritis	Renal Cell Carcinoma
Gross hydronephrosis	Neuroblastoma
Polycystic kidney disease	Renal rhabdoid or clear cell tumour
	Mesoblastic nephroma

Table 2: Differential diagnosis of Wilms' tumour.

Examination and Investigations

The patient presenting with a suspected Wilms' tumour needs careful and thorough examination and investigation. The global health of the child should be considered, height and weight plotted on the growth chart, and routine observations recorded. Of these the most crucial is blood pressure. A full systems examination must be carried out, with notice taken of congenital anomalies, masses and lymphadenopathy. In male patients a varicocoele may be present.

Initial laboratory tests include full blood count, coagulation studies, chemical profile and liver functions. No tumour markers are diagnostic in Wilms' tumours, however many children have a raised LDH. Urinalysis should be performed, and if percutaneous biopsy is not appropriate (see below) urinary catecholamines (HVA, VMA, DOPA) require 24 hour collection to differentiate between Wilms tumour and neuroblastoma.

Imaging

Radiological investigations should be performed and reported by paediatric radiologists experienced in paediatric tumours. Plain X-ray of the abdomen is not necessary, but if performed, calcification is more in keeping with neuroblastoma than Wilms'[8]. A chest X-ray should always be done to look for evidence of lung metastases.

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Abdominal ultrasound is accurate at defining the primary tumour, especially in estimating volume and subsequently, the response to therapy. It is also used to assess for liver metastases, lymph node involvement and peritoneal deposits. It is essential to view the renal veins and inferior vena cava to evaluate for the presence of tumour extension and thrombus. Echocardiography may be required if the thrombus extends in to the right atrium.

Contrast abdominal and thoracic CT is routinely undertaken at the time of presentation for accurate staging. As well as assessing for the presence of metastases, assessment of the contralateral kidney is also undertaken as 5-10% of patients have synchronous bilateral Wilms' tumours. Due to the sensitivity of CT in identifying contralateral tumours, intra-operative assessment is now no longer advised. Following neoadjuvent chemotherapy, routine cross sectional imaging is not required unless there is known vascular tumour extension or preoperative ultrasound raises concerns that require more detailed images to allow operative planning.

MRI scanning is not routinely indicated, but may be useful in situations of intracaval tumour extension and nephroblastomatosis (multiple small renal lesions associated with nephrogenic rests).

Preservation of renal function is vital in cases of bilateral disease. A DMSA scan can be undertaken to plan partial nephrectomy where possible. Selective renal arteriography should also be considered, especially in horseshoe kidneys.

Cardiac toxicity is a common side effect of Doxorubicin; therefore baseline echocardiography should be undertaken in all children and repeated at regular intervals during treatment.

Biopsy

In the UK, an initial clinical and radiological diagnosis of Wilms' tumour is confirmed histologically with a percutaneous biopsy. There are some countries which forgo biopsy and treat presumptively. This is due to fear of tumour rupture or seeding of the biopsy tract, both of which would upstage the tumour. Evidence would suggest that this is unfounded [9,10]. Histology will differentiate other tumour types which require significantly different management. Predominantly cystic tumours and those which have ruptured are not suitable for biopsy.

Treatment

Children with Wilms' tumour present to a variety of healthcare settings initially. It is crucial that where there is suspicion of a renal tumour, the child is referred immediately to a paediatric oncologist or surgeon in order that investigation and treatment is not delayed. A full MDT structure is employed, comprising clinical and radiation oncologists, paediatric surgeons, radiologists, pathologists and specialist nurses. Consideration should be given to inclusion in to an appropriate clinical trial. Such trials further improve our understanding and management of the disease.

Treatment strategy for Wilms' tumour has long divided opinion. Current guidelines differ between the National Wilms' Tumor Study Group (NWTS) and Children's Oncology Group (COG) in North America, the International Society of Paediatric Oncology (SIOP) in Europe and the Children's Cancer and Leukaemia Group (CCLG) in the UK. Event free survival and long term effects are comparable between the groups despite different approaches to investigation and therapy.

Non surgical therapy for Wilms' tumour is guided by radiological or operative stage and histological 'risk'. As both the histology and staging of a tumour can be altered by pre-operative treatment, the staging systems for the UK (table 3) and US are slightly different.

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UK Stage	Description	3 year survival
Stage I	Tumour confined to the kidney and completely resected	90-95%
Stage II	Tumour extends beyond the kidney but is completely resected	80-90%
Stage III Any one of the following	Incomplete tumour resection Tumour in abdominal lymph nodes Tumour rupture (pre or intra-operatively) Peritoneal spread Previous open biopsy	80-85%
Stage VI	Metastatic disease	70-75%
Stage V	Bilateral renal tumours	80-85%

Table 3: UK staging system for Wilms' tumour with corresponding outcomes.

Chemotherapy

Following biopsy, children have 5-7 weeks of pre-operative chemotherapy, the exact regime dependent on their risk stratification. The aim of this is to 'downstage' the tumour, making the surgery easier (smaller mass, less invasion, reduced risk of tumour rupture[11]) and to reduce the burden of therapy required post operatively. Combinations of Vincristine, Actinomycin D, Doxorubicin and various platinum based chemotherapeutic agents are given; details of specific regimes are numerous and beyond the scope of this article.

Surgery

Nephrectomy is planned immediately following chemotherapy for all except those with Stage V disease. Diagnostic doubt, tumour rupture, cystic tumours and age less than 6 months are indications for primary surgery. In the US, nephrectomy is carried out prior to any adjuncts, except in the case of bilateral tumours, where preservation of renal parenchyma is vital. In a small number of children, nephrectomy alone is sufficient[12].

The traditional approach for Wilms' nephrectomy is transperitoneal through a transverse abdominal incision. This allows good access for vascular control, careful dissection and sampling of hilar and para-aortic lymph nodes, in addition to excising the percutaneous biopsy track. The standard principles of tumour surgery apply; careful en bloc resection to avoid rupture and tumour spill, and early control of blood vessels. Ligation of the arterial supply before the venous drainage avoids congestion of the kidney however initial venous ligation may prevent haematogenous spread of tumour cells during handling of the kidney.

As the tumour is sensitive to chemo and radiotherapy, heroic or mutilating resections are not advised.

Partial nephrectomy must be considered in bilateral disease, where the approach is tailored to each child. Partial nephrectomy has been undertaken in unilateral disease, although this is technically feasible in less than 5% of cases [13]. It can be useful in syndromic patients with high risk of metachronous tumour formation, where there is need to preserve renal parenchyma.

The majority of resections can be undertaken in a Paediatric Surgical centre, with a specialist oncological surgeon. For those tumours with significant vascular involvement and thrombus extension well into the vena cava or right atrium, vascular and cardiac surgical support is advised, and cardiac bypass should be considered.

Following nephrectomy, the specimen is sent fresh to the laboratory, and further histological assessment is made. Findings inform the regime of postoperative treatment.

Metastatic disease may be tackled with a combination of chemotherapy, radiotherapy and surgical resection, depending on site and responsiveness to therapy.

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Radiotherapy

Wilms' tumours are radiosensitive; therefore radiotherapy does play a part in the management. Local or whole abdominal irradiation is reserved for those with high risk disease, due to the significant side effects. Non or poorly responsive pulmonary metastases are treated with whole lung irradiation.

Outcomes

Over many years, the outcome for children with Wilms' tumour has improved significantly. This is due to greater knowledge of the pathophysiology, advancements in non-surgical therapy and systematic clinical trials. As seen in table 3, the long term survival (disease free survival at 3 years is equated with cure) for even metastatic disease is very good. Nevertheless, other long term effects must be taken into account. Children receiving treatment for Wilms' tumour have an increased risk of second malignant neoplasms, congestive heart failure, and renal failure [14,15]. Patients are followed up for five years (3-4 monthly for the first three years and six monthly thereafter). Following this, only those who need screening for the late effects of radiotherapy and doxorubicin are reviewed.

Case scenarios

Case 1

This 3 year old girl presented initially to dermatology with enlarged inguinal lymph nodes secondary to infected eczema. Thorough examination at this consultation found bilateral abdominal masses, the left larger than right. She was referred to the paediatric oncology team.

LDH was 703. Contrast CT of her abdomen and thorax found multiple bilateral renal lesions (figure 1) with no enlarged local lymph nodes or vascular invasion. Ultrasound guided percutaneous biopsy was performed of the largest lesion which showed a classical triphasic Wilms' tumour with no high risk features. Biopsy of her inguinal lymph nodes found them to be reactive only.

Management was discussed at the paediatric oncology MDT. A pre-operative regime of vincristine and actinomycin was instituted. After 8 weeks of treatment, the tumours were reassessed with CT (figure 2). 5 months into treatment, she underwent a bilateral renal sparing operation. Further histology proved Wilms' tumour again, but noted nephrogenic rests in the resection margins. Post-operative vincristine and actinomycin chemotherapy continues.

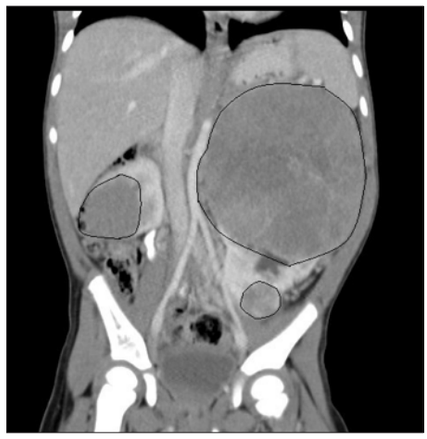

Figure 1: Lines show multiple bilateral Wilms' tumours on a pre-treatment contrast CT

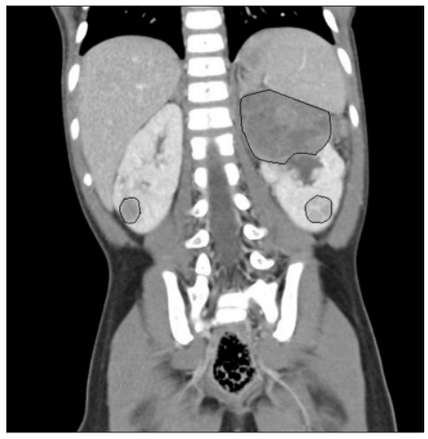

Figure 2: Post chemotherapy CT – lines show reduction in tumour sizes compared to figure 1.

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Case 2

A non-identical twin boy presented aged 5 to his GP with a 2 week history of abdominal distention, pain and loss of appetite. He was lethargic, not managing to keep up with his friends at school, and had lost weight. At first assessment he was found to have a right sided abdominal mass. In addition, he was hypertensive and blood tests showed an LDH of 4488.

Abdominal ultrasound showed a mass arising from the right kidney (figure 3). CT confirmed this (figure 4), and also a single lung met on the right (figure 5). Percutaneous biopsy was carried out, and found a predominantly blastemal type Wilms tumour, with no anaplasia. This, along with stage IV disease at presentation, put him in a high risk category, and therefore underwent 6 weeks pre-operative chemotherapy with actinomycin, vincristine and doxorubicin. Pre-operative CT showed a reduction in tumour size (figure 6), and that the lung nodule had altered, likely replaced with scar tissue. He underwent nephrectomy, at which the tumour was found to be adherent to the liver and inferior vena cava. Histology showed extensive chemotherapy response and necrotic involved lymph nodes. 28 weeks of triple chemotherapy are planned post-operatively. The remaining lung nodule may require surgical excision.

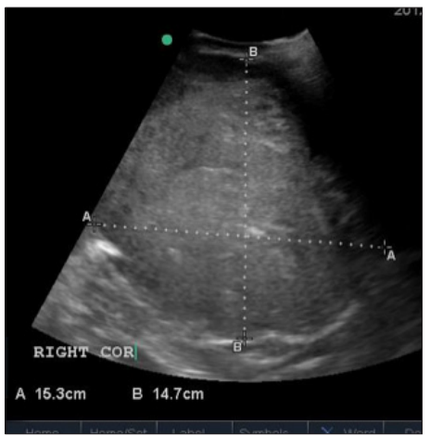

Figure 3: Ultrasound scan showing a mass arising from the right kidney.

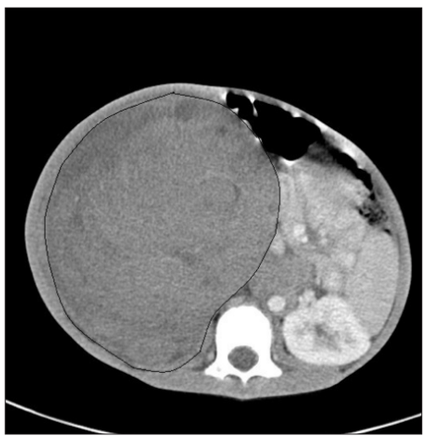

Figure 4: Pre-treatment CT showing a right sided Wilms' tumour (line)

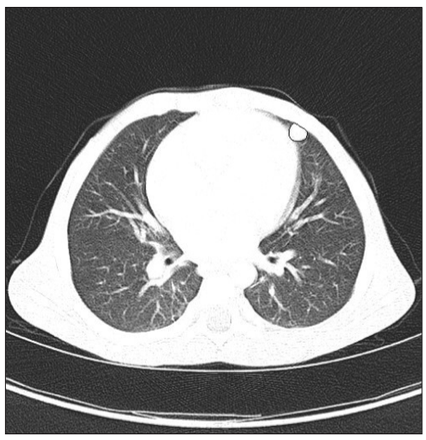

Figure 5: Pre-treatment CT thorax with lung windows showing a solitary lung metastasis (line)

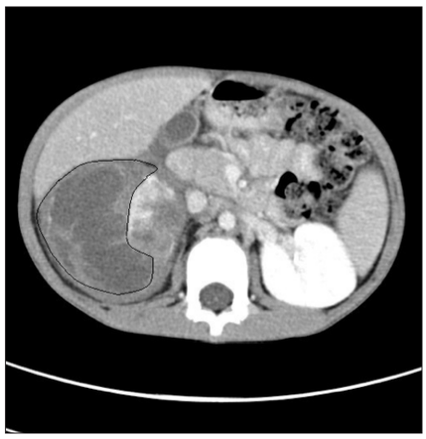

Figure 6: Post chemotherapy contrast CT showing reduction in tumour size compared to figure 4.

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Multiple choice questions

A. Wilms' tumour

1. Frequently occurs <6 months of age	T	F
2. Is associated with very low birth weight	T	F
3. At presentation is frequently metastatic	T	F
4. Enjoys a generally good prognosis	T	F
5. Frequently presents with abdominal pain	T	F

B. The following conditions are associated with Wilms' tumour

1. Beckwith Weidemann Syndrome	T	F
2. Downs Syndrome	T	F
3. Denys Drash Syndrome	T	F
4. AAA syndrome	T	F
5. Prematurity	T	F

C. The following are indicators of a poor prognosis in Wilms' tumour

1. Loss of heterozygosity 16q	T	F
2. WT1 gene	T	F
3. Loss of heterozygosity 1p	T	F
4. MYCN amplification	T	F
5. unbalanced 11p aberration	T	F

D. Regarding Histopathology

1. Wilms' tumour is biphasic	T	F
2. 5% of Wilms' tumours are associated with nephrogenic rests	T	F
3. Necrotic tissue at diagnosis is a poor prognostic sign	T	F
4. Wilms tumours can contain calcification	T	F
5. Are bilateral 10% time	T	F

E. Investigation and treatment for Wilms' tumour:

1. Includes initial nephrectomy in all cases	T	F
2. Wilms' tumours are radiosensitive so this forms a core therapy in all bar stage 1 tumours	T	F
3. Pre-operative percutaneous biopsy upstages the tumour (increases the risk of rupture and seeding)	T	F
4. DMSA is a routine preoperative investigation	T	F
5. Calcification on an x-ray confirms the diagnosis of Wilms' tumour	T	F

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Answers

A. 1)F 2)F 3)F 4)T 5)F

B. 1)T 2)F 3)T 4)F 5)F

C. 1)T 2)F 3)T 4)F 5)F

D. 1)F 2)F 3)F 4)F 5)T

E. 1)F 2)F 3)F 4)F 5)F

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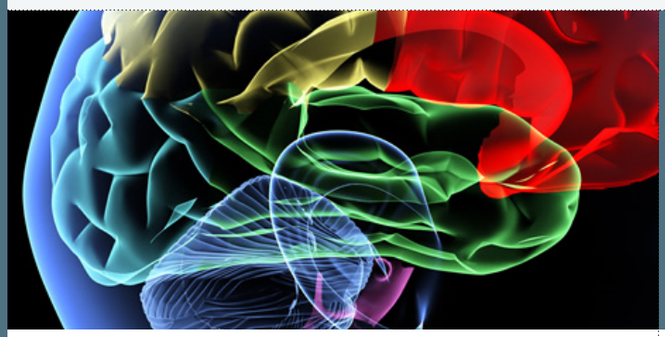
GLIOMAS

A Roylance, MS Palin, A Zafar



Gliomas
Neurosurgery

Abstract

Gliomas are the most common type of primary brain tumour. They arise from glial cells, of which there are different types with varying functions. Types of glioma include astrocytoma, oligodendroglioma and glioblastoma multiforme. The degree of malignancy varies, as does corresponding radiological and histological features. Gliomas affect both adult and paediatric populations with increased incidence of infratentorial neoplasms in children. There is a number of treatment options available ranging from conservative management to radical resection with multimodality radiotherapy. Stereotactic radiosurgery is used for smaller, lower grade deep cortical lesions. Surgical resection is preferred for anaplastic lesions with associated oedema and increased vascularity such as glioblastoma.

Keywords: *Glioma, Brain Tumour, Neoplasm, Histology, Imaging.*

Introduction

Gliomas are a type of intrinsic brain tumour arising from neuroglial cells and account for approximately eighty percent of all malignant primary brain tumours (1). There are various types of neuroglial cell with roles ranging from providing intrinsic support to neurons, to facilitating production of myelin. Glial cells also exhibit functionality in neurotransmitter regulation and modulating synaptic homeostasis (2).

Glia may also be categorised as either micro or macroglia. Microglial cells are primarily associated with phagcytosis whilst macroglia can be categorised into further subtypes, each having distinct functions. These include astrocytes, oligodendrocytes and ependymocytes (ependymal cells) (3). Less abundant subtypes include radial and satellite glia which are associated with neuronal progenation and chemical homeostasis respectively (4).

Astrocytes are the most common type of neuroglial cell. They are also involved in chemical homeostasis as well as regulating neurotransmitter regeneration and vasoactivity via arachidonic acid synthesis (5). Oligodendrocytes primarily manufacture myelin, a lipoprotein that insulates neuronal axons and augments action potential propagation (6). Ependymocytes help form the walls of the ventricular system and are involved in secretion and circulation of cerebrospinal fluid (CSF).

Types of glioma

Different types of glioma exist in conjunction with such cell types, the most common being astrocytoma. Astrocytomas account for fifty percent of all primary brain tumours (7). They are typically supratentorial but have a relatively increased cerebellar incidence in children. Glioblastoma multiforme is the most common type of astrocytoma, a highly malignant sporadic variant with no identified genetic predisposition unless concomitant with disorders such as Von-Hippel Lindau, Li Fraumeni and neurofibromatosis (8). It affects more males, has been linked with increased alcohol consumption, ionising radiation and certain viruses (9). It has an average incidence of 2.5 per every 100,000. Due to their capacity for rapid growth, the most commonly associated symptoms are those of raised intracranial pressure. These include nausea and vomiting, decreased conscious level and headache. However, a patient may develop specific symptoms based on the exact location of the tumour such as visual disturbances, aphasia or motor impairment.

Oligondendrogliomas are the second most common brain tumour in adults accounting for four percent of all primary brain tumours (7). They arise from oligodendrocytes and are mostly located in the frontal and temporal lobes. The pathophysiology of oligodendrogliomas is unclear although recent literature suggests characteristic losses at chromosomes 1p and 19q (10). Oligondendrogliomas are more common in young and middle aged adults and are often associated with seizures, headaches or behavioural changes in keeping with their typical frontotemporal distribution (11).

Ependymomas are a type of glioma arising from ependymal cells. They commonly evolve within the ventricular system in adults and are often associated with disruption of CSF and hydrocephalus. They are relatively more common in the cerebellum amongst infants. Ependymomas account for two to three percent of all primary brain tumours, although the incidence in paediatric patients is approximately ten percent with a third of patients being three years or younger. Peak incidence in adults is approximately thirty five years of age. Ependymomas are typically divided into four subtypes. These are subependymomas, myxopapillary, anaplastic and 'non-specific' ependymomas which themselves can be classified as cellular, papillary, clear cell or tancytic. The vast majority of ependymomas are myxopapillary in nature which are benign, slow growing low grade tumours. However, anaplastic ependymomas are characterised by rapid growth (12).

GLIOMAS

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Pleomorphic xanthoastrocytomas are usually low grade lesions which tend to arise in children and young adults with a prolonged history of seizures. They are commonly seen in superficial cortical areas, in the temporal lobes and frequently involve the leptomeninges. Maximal surgical resection is the treatment of choice where possible, with long term survival rates of approximately ninety percent.

WHO Classification	Example	Possible histological signs	Median survival
I	Pilocytic Astrocytoma	Increased cellularity	8-10 years
II	Fibrillary Astrocytoma	Cytological atypia	7-8 years
III	Anaplastic Astrocytomas	Anaplasia and mitotic activity	2-3 years
IV	Glioblastoma Multiforme	Microvascular proliferation +/- necrosis	<1 year

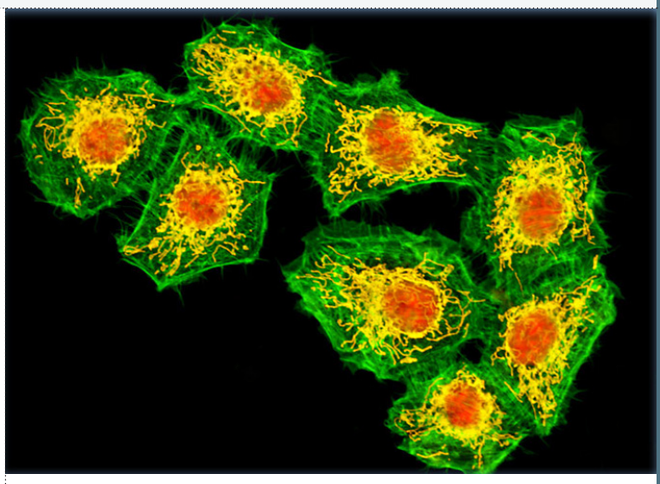
Table 1: WHO glioma classification, corresponding histological findings and anticipated median survival time.

Histological Assessment

There are various grading systems for staging astrocytomas. The World Health Organisation (WHO) system is the most commonly used and contemporary method. It is a three tiered system in which non-pilocytic astrocytomas (astrocytic tumours) are subdivided into low grade (fibrillary) astrocytomas, anaplastic astrocytomas and glioblastoma multiforme based on the degree of anaplasia. Tumours are given a numeric grade reflecting the number of histological criteria they exhibit (13).

Grade II (fibrillary) astrocytomas for example, display a predominance of astrocytes without clear anaplasia, vascular proliferation or necrosis. Grade III lesions (anaplastic astrocytomas) exhibit focal anaplasia such as hypercellularity, nuclear pleomorphism or vascular proliferation (14). Glioblastoma multiforme, the most anaplastic lesion (Grade IV) exhibits more widespread anaplastic cells often accompanied by glial processes and significant histological haemorrhage in addition to the pathological features of lower grade lesions (15). Grade I tumours (pilocytic astrocytomas) may demonstrate increased cellularity but possess otherwise normal cytology. They mostly occur in the cerebellum, cerebrum, optic nerve pathway and brainstem (16).

Pleomorphic xanthoastrocytomas act in a far less aggressive fashion than other pleomorphic gliomas despite high cellular activity. They exhibit minimal mitotic activity or necrosis but demonstrate nuclear hyperchromasia and consist of spindle cells arranged into fascicles, intersecting bundles or a storiform pattern. Perivascular lymphocytic collections as well as eosinophilic granular bodies (EGBs) are common. Giant xanthomatous cells with intercytoplasmic lipid droplets are less common but an accurate diagnostic indicator (17).



Oligodendrogliomas possess uniform cellularity with oval nuclei and well defined cell membranes. These lesions are associated with a distinctive 'fried egg' appearance due to the formation of perinuclear halos following tissue processing. There may be signs of anaplasia in anaplastic oligodendrogliomas or evidence of astrocytic cells in 'mixed' lesions (10).

Ependymomas have characteristic cell organisation with ependymal cells forming uniform patterns of rosettes, perivascular pseudorosettes or canals surrounded by cuboidal cells. Small dark nuclei are seen and anaplastic cell changes may occur in some cases with an increased incidence in cerebral and fourth ventricular lesions (12).

Imaging

Although histological assessment is still the definitive modality for unequivocal staging of gliomas, neuroimaging may be used to augment diagnosis. Both CT and MRI are used to assess factors such as calcification, vasogenic oedema, necrosis, cyst formation and lesion enhancement. However, recent research suggests the efficacy of such modalities is limited. Conversely, the use of other parameters such as metabolite ratios and relative cerebral volume in conjunction with more contemporary imaging techniques such as diffusion tension imaging (DTI) tractography and functional magnetic resonance imaging (fMRI) is suggested. Recurrence may also be predicted relatively early due to metabolite changes observable via positron emission tomography (PET).

Characteristic findings

Pilocytic astrocytomas appear hypodense to isodense on CT with a macrocystic hypodense core in half of cases. Calcification is seen in approximately twenty percent and fourth ventricular compression is also an anticipated radiological sign. On MRI, a pilocytic astrocytoma will appear as a sharply defined macrocystic mass with prolonged T1 and T2 relaxation times. A mural nodule is often identifiable due to its pronounced contrast enhancement (18).

GLIOMAS

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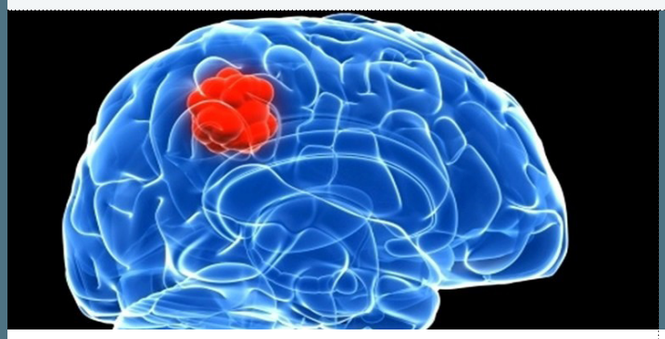

Gliomas
Neurosurgery

Grade II astrocytomas are typically hypodense on CT with negligible oedema or mass effect. They are less contrast enhancing than other types of glioma and their appearance on MRI is similar to that of pilocytic astrocytoma with a well defined lesion and prolonged relaxation times. Conversely, anaplastic astrocytomas exhibit less defined margins and produce a more heterogeneous signal when compared to less anaplastic lesions. The presence of associated oedema and mass effect is common with contrast enhancement in most cases (18).

Characteristic irregular areas of calcification are associated with oligodendroglioma. These may be seen on CT or MRI and manifest as areas of low attenuation. Cystic changes and haemorrhage are infrequently observed and as with anaplastic and pilocytic lesions, tissue exhibits hypo/isodensity (11), (18).

Ependymal tumours display mixed density with fine calcification in approximately half of cases. Enhancement is common and may also be associated with cystic areas, particularly in the cerebral hemispheres (12), (18).

Glioblastoma multiforme typically causes significant oedema and mass effect on CT and MRI. Margins are poorly defined but more than 95% of tumours will enhance post contrast with irregular annular enhancement. Glioblastomas may also be seen to cross the midline and cause haemorrhagic changes (18).

Type	Treatment
Pilocytic Astrocytoma	• Maximal surgical resection with serial CT/ MRI post operative surveillance
Fibrillary Astrocytoma	• Serial neurological examination and imaging • Radiotherapy • Chemotherapy • Chemotherapy and Radiotherapy • Surgery
Anaplastic Astrocytomas Glioblastoma Multiforme	• Cytoreductive surgery and subsequent external beam radiation • Chemotherapy: i.e. Alkylating agents, implantable agents (Gliadel wafers)

Table 2: Varying tumour types and recommended treatments.

Epidemiology

Pilocytic astrocytomas account for approximately 2% of all brain tumors. They most commonly present in children and adolescents and are often referred to as Juvenile Pilocytic Astrocytoma (JPA) as a result (17).

WHO grade II astrocytomas typically arise in the third to fifth decades, are slow growing and associated with seizures. Anaplastic astrocytomas however, most commonly present in the fifth and sixth decades of life. They are typically supratentorial and exhibit symptoms such as seizures and focal neurological deficits (18).

The incidence of glioblastoma increases with age, with peak prevalence in the sixth and seventh decades. However, GBM may occur at any age and typically affects the cerebral hemispheres and corpus callosum. They rarely involve the deep grey matter, brainstem and cerebellum and are not usually associated with seizure activity (18).

The incidence of oligodendrogliomas is thought to be increasing due to improvements in staining techniques. However, as with anaplastic astrocytomas, peak incidence is still anticipated to be in the fifth and sixth decades of life.

Ependymomas have a relatively high incidence in children and are the third most common paediatric tumour. Infratentorial lesions are more common in this population where as supratentorial ependymomas are more common in adult patients (17).

Treatment

Interventional criteria

Surgical management is indicated in the majority of glioma patients. As oppose to solely offering a definitive cure, surgical intervention is used for symptom control and tissue biopsy apart from in paediatric brainstem lesions. In actuality, apart from pilocytic astrocytomas, pleomorphic xanthroastrocytomas and the minority of well circumscribed low grade gliomas, most lesions cannot be cured with surgery alone. The degree of aggressive intervention correlates with histological outcome with interventions ranging from periodic surveillance scans to radical resection with multimodality radiotherapy.

GLIOMAS

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Symptoms are largely associated with mass effect from the glioma itself, haemorrhage or surrounding oedema. Recent literature suggests effective relief of raised intracranial pressure and hydrocephalus via cytoreduction of neoplastic cells. Maximal safe cytoreduction also substantially reduces the chance of future malignant degeneration, the development of seizures as well as the reliance on anti-epileptics and steroids. Surgical de-bulking also enhances a patient's tolerance of adjuvant therapies and yields higher Karnofsky scores (18), (19).

When assessing a patient's suitability for surgery as well as the type of surgical intervention that is most appropriate, be it stereotactic or craniotomy, a number of factors should be considered. These can be divided into patient or tumour related factors. When assessing the lesion itself, its size, degree of mass effect and vascularity should be evaluated. Composition (solid or cystic) and location are other important factors, with particular reference to whether a tumour occupies eloquent or silent brain areas.

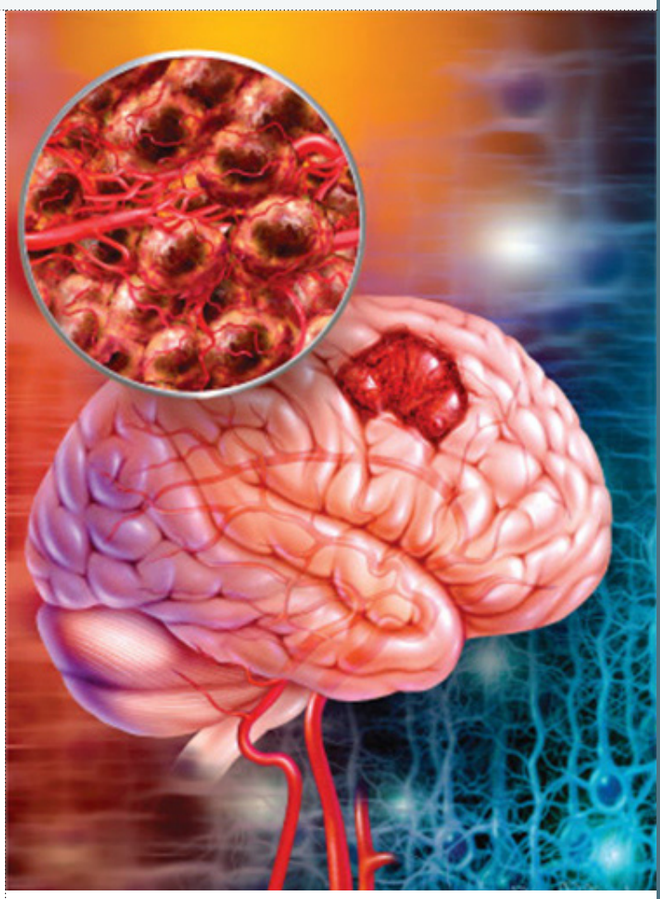
Patient related factors include neurological status (Karnofsky score), prior therapy, patient/family wishes and surgical risks such as bleeding, infection or delayed healing. These will be influenced by more generic factors including patient age and co-morbidities. It is proposed that survival rates are doubled if a patient's Karnofsky is seventy or above (19). With regards prior therapy, side effects of chemotherapy including bone marrow suppression and anaemias are to be considered (18).

Regarding diagnostic use, stereotactic biopsy is suggested for non-resectable tumours to facilitate histology guided adjuvant therapy. However, tumour sampling via craniotomy is widely considered to reduce sampling errors otherwise experienced through stereotactic means. The choice to proceed with closed stereotactic biopsy, open biopsy or surgical resection is based on various factors (20).

Surgery

Biopsy

Stereotactic biopsy is favoured in patients exhibiting small deep lesions with a unilocular cystic component generating minimal mass effect. It is also preferred in older, neurologically intact patients or those with higher medical and anaesthetic risk such as bleeding diathesis. The diagnostic accuracy of stereotactic sampling is less than surgical resection with values of 72 to 93%. Stereotactic intervention should also be avoided in highly vascular lesions. In this instance, open biopsy or surgical resection are the preferred approaches (18). Open methods are also indicated for lesions in eloquent brain areas requiring cortical mapping. Surgical resection is favoured in lower risk patients, those exhibiting neurological impairment and for large, superficial loculated lesions (21).



Surgical techniques

There are four routine types of craniotomy utilised in glioma surgery including pterional, Sutar (bicoronal), horseshoe and sub-occipital approaches. Temporal and frontal tumours proximal to the Sylvian fissure and skullbase can be accessed via a pterional approach. This can be extended posteriorly or towards the vertex to access the tempoparietal and lateral frontal regions. The desired incision spans in a 'question mark' shape from the level of the zygomatic process posterior to the palpable pulse of the superficial temporal artery to slightly anteromedial of Kocher's point. Care must be taken to avoid incisional injury to the frontotemporal branch of the facial nerve (21).

For frontal lesions too anteromedial for a pterional craniotomy, a bicoronal incision is used. Access to the chiasm and anterior hypothalamic region is also possible by a subfrontal variant. An incision is made between the zygomatic processes usually 1cm posterior to the hairline (18). For a unilateral approach however, the incision is made to the contralateral superior temporal line. A skin flap is deflected in the supraperiosteal plane with avoidance of the superficial temporal artery and terminal branch of the facial nerve (supraorbital) as it exits the supraorbital foramen and provides palpebral filaments supplying the conjunctiva and eyelids. Care should also be taken to avoid compromising the frontal air sinus and the development of a secondary fistula with the intradural space (18), (21).

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Gliomas
Neurosurgery

The intraventricular region and interhemispheric areas such as the corpus callosum and interhemispheric fissure can be accessed by a lateral horseshoe craniotomy (18). If made adjacent to the coronal suture, the anterior aspect of the third ventricle can be accessed transcallosally through the Foramen of Monroe. This approach is preferred for midline frontal lobe gliomas, hypothalamic astrocytomas and ependymal cell lesions proximal to the lateral ventricles. However parietal and occipital lobe tumours can be accessed by a more posterior horseshoe opening. Care should be taken so as to not damage the superior sagittal sinus.

Infratentorial lesions such as cerebellar astrocytomas or ependymomas can be accessed via a sub-occipital craniotomy in either a midline or paramedian position (17). For a midline approach, an incision is made from the inion to the second cervical spinous process. Overlying musculature is dissected and reflected in a subperiostal plane (18). Preservation of the vertebral arteries in the region of the foramen magnum is essential. As well as the foramen magnum, the boundaries of the sub-occipital approach are the transverse sinuses rostrally and the transverse-sigmoid junctions laterally. For more lateral lesions, linear or sigmoidal incisions can be utilised (21).

Cortical mapping

Various cortical mapping methods can be used to maximize tumour resection whilst preserving normal brain. These include generating 3D schematics to aid peri-operative navigation, electrophysiological testing and the use of functional imaging modalities such as functional MRI and DTI tractography (22). These may be used to map eloquent cortex such as speech and language areas preoperatively or during awake craniotomy. The use of electrophysiological testing in such patients is also increasingly favoured with developments in direct cortical stimulation, negative mapping and subcortical stimulation.

Direct cortical stimulation can be used in various brain areas to determine eloquent cortex. By doing so, safe resection margins can be devised. However, this method typically requires an extensive craniotomy. Conversely, negative mapping relies on localization of cortical regions that, when tested, contain no stimulation-induced functional change. This has led to less-extensive intraoperative mapping, smaller craniotomy and a more time-efficient neurosurgical procedure (22).

Multiple choice questions

Please select the single most appropriate response:

1. Gliomas account for what percentage of malignant primary brain tumours?

a) 20%
b) 40%
c) 60%
d) 70%
e) 80%

2. Regarding radiological assessment

a) Oligodendroglioma most commonly arise in the parietal and occipital lobes
b) Glioblastomas do not cross the midline
c) A macrocystic hypodense core is a feature of pilocytic astrocytomas
d) Areas of irregular calcification are associated with pleomorphic xanthoastrocytomas on CT scan
e) Glioblastomas rarely respond to contrast medium

3. Regarding histological evaluation

a) Ependymomas are characterized by the formation of rosettes and pseudorosettes on microscopy
b) Fibrillary astrocytomas exhibit a characteristic 'fried egg' appearance on microscopy
c) Intercytoplasmic lipid droplets are defining feature of ependymal cell tumours
d) Anaplastic astrocytomas are the most neoplastic type of glioma
e) Oligodendrogliomas exhibit glial processes and significant haemorrhage on histological analysis

4. Epidemiology

a) Anaplastic astrocytomas typically present in teenage years
b) The incidence of glioblastoma decreases with age
c) Supratentorial ependymomas are most common in paediatric patients
d) WHO grade II astrocytomas typically arise in the third to fifth decades
e) Pilocytic astrocytomas most commonly occur in adults

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5. Surgical treatments

a) Surgical biopsy in encouraged for paediatric brainstem lesion
b) Stereotactic tissue sampling produces more accurate results than resection
c) Stereotactic surgery is indicated in highly vascular lesions
d) Care should be taken to avoid the supraorbital nerve when performing a unilateral anterolateral craniotomy
e) Bicoronal craniotomy is the preferred method for midline lesion

Answers

1. E
2. C
3. A
4. D
5. B

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REVALIDATION – ARE YOU READY?

J Risley

Abstract

Every doctor will have to undergo revalidation at least once in their career. It does, however, remain an issue that to many may not be clear. The importance to engage with the process, no matter what the stage of training of the individual doctor, should not be underestimated, and indeed doing so early will make the process significantly less stressful and time consuming.

Keywords: *Revalidation; General Medical Council; Good Medical Practice; Appraisal; Licence to practice.*

The aim of this article is to briefly outline the process of revalidation, and to explain what will be required of you during that process. There is a wealth of information on the GMC website, and I would direct your attention to the documents entitled 'Ready for revalidation: The Good Medical Practice framework for appraisal and revalidation 2012' (1), and 'Ready for revalidation: Supporting information for appraisal and revalidation 2012' (2) on which this article is based.

Revalidation commenced on the 3rd of December 2012, with the GMC expecting to revalidate the majority of licensed doctors by March 2016. By holding a license to practice you are legally required to revalidate, usually every five years, by having regular appraisals based on the core guidance for doctors, Good Medical Practice.

In order to continue to hold a license and practice, you must successfully revalidate. The aim of revalidation is to give patients greater confidence that you are up to date in the area of medicine in which you practice. It also aims to support you in maintaining and developing your practice throughout your career in medicine, by ensuring that you have the opportunity to regularly reflect on how you can change and improve your practice.

Preparation for revalidation

You will not be able to revalidate without having a regular appraisal; this should not be a problem for those in training posts, but for those that take time out for research, personal reasons, career breaks, or those in locum positions, it is important for you to bear this in mind. The appraisal must be based on the principles as outlined in Good Medical Practice.

You are responsible for taking reasonable steps to collect the information required for your appraisal, however the organization that you work for (e.g. your Trust) should help you by giving you access to the information that you need, such as complaints and compliments.

You should set up your online GMC account, which will provide all of your revalidation details such as the name of your responsible officer, and your designated body.

Your responsible officer is the person who will make a recommendation to the GMC to revalidate you, and is a licensed doctor who is usually the medical director or their deputy of that organization. They will act on behalf of your designated body, which is the organization that has a duty to provide you with a regular appraisal and support you with revalidation.

It is important to keep your designated body information up to date via GMC online. If you are new to an organization, you should not assume that the responsible officer has added you to their list for revalidation, and you should update your designated body information yourself on GMC online. The system will then alert the relevant responsible officer that you have connected to their organization.

REVALIDATION – ARE YOU READY?

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The revalidation submission date is the date by which the GMC need to receive a recommendation about you from your responsible officer. They may make one of three recommendations about you:

· They can make a positive recommendation that you are up to date, fit to practice, and should be revalidated.

· Request a deferral because they need more time or more information to make a recommendation about you. This might happen if you take an extended break from practice. Deferral does not affect your license to practice.

· Notify the GMC that you have failed to engage with appraisal or any other local systems or processes that support revalidation.

The GMC will then consider the recommendation, and contact you with their decision as to your revalidation.

GMP Framework for appraisal and revalidation

This is based on Good Medical Practice. The framework (1) sets out the broad areas which should be covered in a doctor's appraisal, and on which the recommendations to revalidate a doctor should be based. The framework can allow doctors to:

- Reflect on their practice and their approach to medicine
- Reflect on the supporting information they have gathered and what that information demonstrates about their practice.
- Identify areas of practice where they could make improvements or undertake further development.
- Demonstrate that they are up to date and fit to practice.

The framework consists of four domains that cover the spectrum of medical practice. In addition, each of the domains is described by three attributes. You need to maintain a portfolio of supporting information to demonstrate that you are continuing to meet the below:

• Domain 1 – Knowledge, skills and performance
- 1.1 Maintain your professional performance
- 1.2 Apply knowledge and experience to practice
- 1.3 Ensure that all documentation formally recording your work is clear, accurate and legible.

• Domain 2 – Safety and quality
- 2.1 Contribute to and comply with systems to protect patients
- 2.2 Respond to risks and safety
- 2.3 Protect patients and colleagues from any risk posed by your health.

• Domain 3 – Communication, partnership and teamwork
- 3.1 Communicate effectively
- 3.2 Work constructively with colleagues and delegate effectively
- 3.3 Establish and maintain partnerships with patients

• Domain 4 – Maintaining trust
- 4.1 Show respect for patients
- 4.2 Treat patients and colleagues fairly and without discrimination
- 4.3 Act with honesty and integrity

Supporting information for appraisal & revalidation

This guidance document (2) sets out the supporting information that you will need to provide at your annual appraisal and the frequency with which it should be provided. It falls under four broad headings:

1. General information – providing context about what you do in all aspects of your work.

2. Keeping up to date – maintaining and enhancing the quality of your professional work.

3. Review of your practice – evaluating the quality of your professional work.

4. Feedback on your practice – how others perceive the quality of your professional work.

REVALIDATION – ARE YOU READY?

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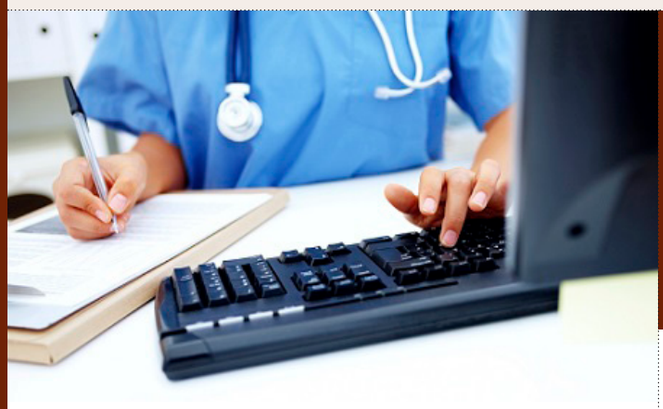

There are six types of supporting information that you will be expected to provide and discuss at your appraisal at least once in each five year cycle. They are:

1. Continuing Professional Development.

2. Quality improvement activity

3. Significant events

4. Feedback from colleagues

5. Feedback from patients.

6. Review of complaints and compliments.

By providing all six types of supporting information over the revalidation cycle you should, through reflection and discussion at appraisal, have demonstrated your practice against all twelve attributes outlined in Good Medical Practice Framework for Appraisal and Revalidation. This will make it easier for your appraiser to complete your appraisal and for your Responsible Officer to make a recommendation to the GMC about your revalidation.

Revalidation – are you ready?
Current Training Issues

1. Continuing Professional Development (CPD)
This should encourage and support specific changes in practice and career development, and be relevant to your practice. There is no formal amount of CPD that is required for revalidation, though the Royal Colleges may provide a guide depending on your specialty. This would be approximately 50 hours per year, totaling 250 hours over a revalidation cycle (3).

2. Quality Improvement Activity
You will have to demonstrate that you regularly participate in activities that review and evaluate the quality of your work. Examples include clinical audit and case-based discussions. It is important that you have evaluated and reflected on the results of the activity or audit.

3. Significant events
Also known as an untoward or critical incident; it is any unintended or unexpected event, which could or did lead to harm of one or more patients. Again these should be identified and reflected upon.

4. Colleague feedback and 5. Patient feedback
The GMC has developed colleague and patient questionnaires that any doctor can use, though it is expected that any questionnaire will be administered independently of the doctor and the appraiser. There are however third party companies which can provide such a service. With regards patient feedback, the GMC has not prescribed the number of patient responses that you are required to collect, however it is thought approximately 35 would suffice.

5. Review of complaints and compliments
A complaint should be seen as another type of feedback, allowing doctors and organizations to review and further develop their practice to make patient-centred improvements.

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Conclusion

Revalidation should not pose many problems for those in formal training schemes; many of the requirements for revalidation will be the same, if not similar, to those required for a trainee to successfully progress through specialty training. Trainees should however bear in mind the requirement for patient feedback, the importance of reflecting upon evidence provided at appraisal, and the importance of ensuring adequate CPD. For those not in training posts, it is likely that the process will not be as straightforward; the importance of a portfolio and taking a proactive approach to the process should not be underestimated.

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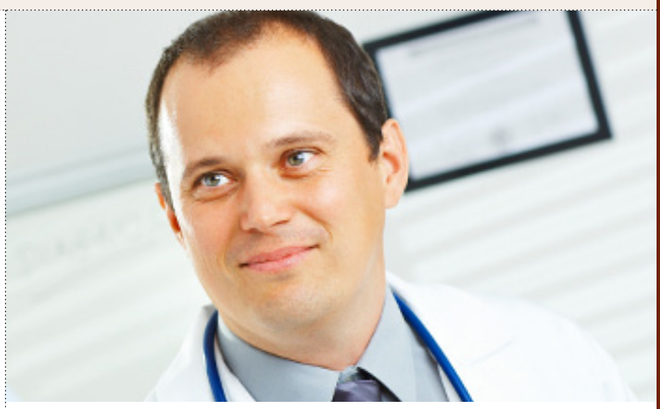

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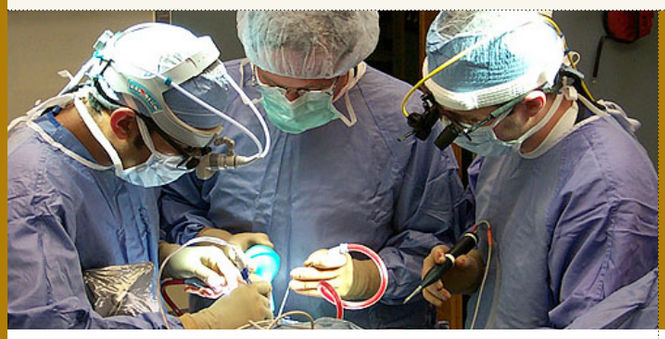
COURSE REVIEW - CAMBRIDGE LECTURES IN NEUROSURGICAL ANATOMY

D Fitzrol



Course Review - Cambridge Lectures in Neurosurgical Anatomy
Career Focus

Abstract

Courses are increasingly important for ongoing medical education in all medical and surgical specialities, particularly with the reduction of working hours due to the European Working Time Directives (EWTD) resulting in less hands-on experience for trainees. Neurosurgery is not exempted from this and the importance of neuroanatomy as the foundation of neurosurgery cannot be emphasised enough. Cambridge Lectures in Neurosurgical Anatomy is a course targeted to those who are keen on either to learn or consolidate knowledge on neuroanatomy and its relevance in clinical and operative neurosurgery. This course review provides more information about the course including its delegates, structure, fees and what was like to attend the course.

Keywords: *Neurosurgical Anatomy, neurosurgery, course.*

Introduction

Often the passion for neuroanatomy and surgery forms the basis for career pursuit in neurosurgery. The intricacy of the nervous system appeals to the inquisitive mind and challenges one's skills in the operating theatre. Neurosurgery itself is a rapidly advancing field with various techniques being developed. Hence it is essential for the knowledge of neuroanatomy and neurosurgical skills to be consolidated and updated as any mishaps during surgery can lead to devastating consequences. Cambridge Lectures in Neurosurgical Anatomy, established in 2005, serves to provide a pleasant environment where the knowledge of operative neuroanatomy and neurosurgical techniques can be shared and appreciated amongst enthusiastic neurosurgeons and trainees.

What is it?

Cambridge Lectures in Neurosurgical Anatomy is a course whereby lectures on relevant neurosurgical anatomy are carried out in three dimensional (3-D) views of cadaveric dissections by qualified neurosurgeons from Addenbrooke's Hospital, Cambridge. This course is held twice a year in one of the colleges of the University of Cambridge. Each meeting has its own theme on which discussion topics and the teachings of neuroanatomy will be based on. There is also a guest speaker present for each meeting who is world renowned for his subspecialty which also corresponds with the theme of the meeting. The guest speaker will present on topics consisting of a combination of surgical techniques, recent studies, interesting case studies and advances relevant to his area of interest. This allows discussions on various neurosurgical topics and sharing of knowledge between neurosurgeons in the UK and overseas. When I attended the course in December 2012, it was held in Peterhouse College and the meeting then focused on intrinsic brain tumours and epilepsy.

Who is it for?

The course is aimed at neurosurgical trainees and qualified neurosurgeons in the UK and overseas. Junior trainees and medical students who are keen in pursuing a career in neurosurgery or have a strong interest in neuroanatomy are also welcome to attend the course. For junior trainees, this course helps to expand their knowledge and for more senior trainees, this course helps to consolidate and refresh their neurosurgical anatomy. For every delegate, there is always something new to learn about neurosurgery.

How is the course structured?

The course generally runs over two days on a weekend usually in June and in December. It is lecture-based with opportunities to ask questions and discuss different opinions. The day normally begins with a few sessions of neuroanatomy followed by topics relevant to the theme which are presented by the guest speaker. In December 2012, the anatomy lectures were on anterior, middle and posterior skull base with the relevant surgical approaches followed by discussions on meningioma.

These lectures are divided with breaks in between to allow further discussions or to have a chance to rest and socialise with other delegates. Although it is lecture-based, it can be tiring especially for a junior trainee when there is a lot of new information to take in. So, these breaks are very helpful. There is also a session for group photo on Saturdays during the lunch break creating a feeling of camaraderie.

The photos are made available on the website for download after the course. One of the highlights of the course, other than being able to meet some of the world-class experts in neurosurgery is the course dinner organised on Saturdays. It is usually held in the dining hall of the same college where the course is running. It is a 3-course dinner with traditional English food served and the ambience of the college gives a very relaxing and calming effect after a long day.

COURSE REVIEW - CAMBRIDGE LECTURES IN NEUROSURGICAL ANATOMY

D Fitzrol

Are there exams?

No. There are no exams as the course is solely for learning purposes for its delegates.

How much does it costs?

The cost varies but generally about £200 or more. When I attended the course in 2012 it was £260.00. There is no extra charge for the course dinner. Accommodation is not included in the fees.

What is it like to attend?

I attended the course as a Foundation Year 2 trainee. The neuroanatomy lectures can be intense as there is a lot of new information to learn in a short period of time. I found doing some background reading on the neuroanatomy topics in advance helped me understand and follow the lectures much better. It is essential to understand these lectures as they complement the topics on neurosurgical techniques which take place afterwards. Without knowing the relevant operative anatomy, it is difficult to appreciate what is being discussed about the surgical techniques.

This course has also helped me recognise anatomical structures better in operating theatres as it included surgical videos which helped me familiarise myself with both the surgical technique and the relevant anatomy. Moreover, as a junior trainee, listening to the discussions and experiences of these well-established neurosurgeons has been very inspirational and emphasised the need for continuous learning in the field of neurosurgery.

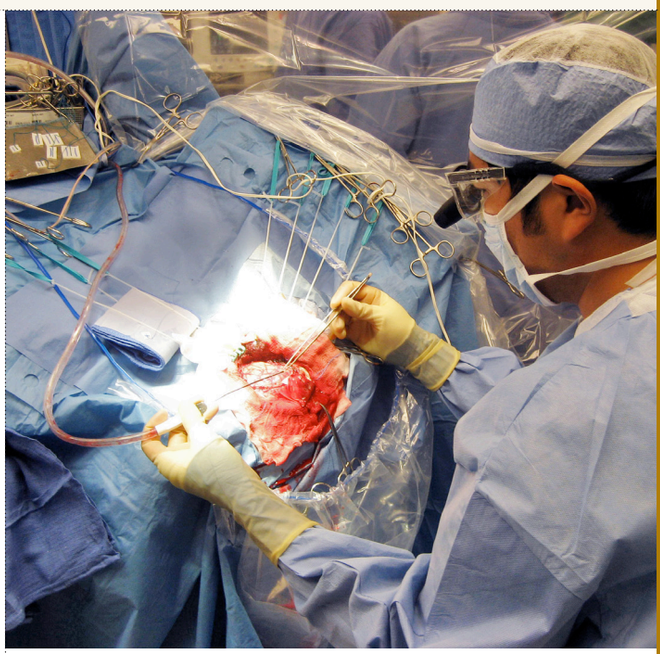
This course is useful for junior trainees who are interested in a more detailed grasp on neurosurgical anatomy and operative techniques. However, it does require good background knowledge prior to attending this course to be able to fully benefit from it, so I would strongly advise preparing for it.

Was it worth it?

Yes, I think it is worth it. Before having the chance to work in a neurosurgical department, this course was where I got a good overview of neuroanatomy, recognising different types of surgical techniques and increasing my knowledge of the latest research studies in the specialty. On top of that, I am more aware of subspecialties that require further research to increase our knowledge base. Besides that, since each meeting has a different theme, delegates can choose which meeting to attend depending on their area of interest. It is also a good chance to learn on how neurosurgery is practised in different countries from the guest speaker.

Links for more information:

http://www.clna.org.uk/



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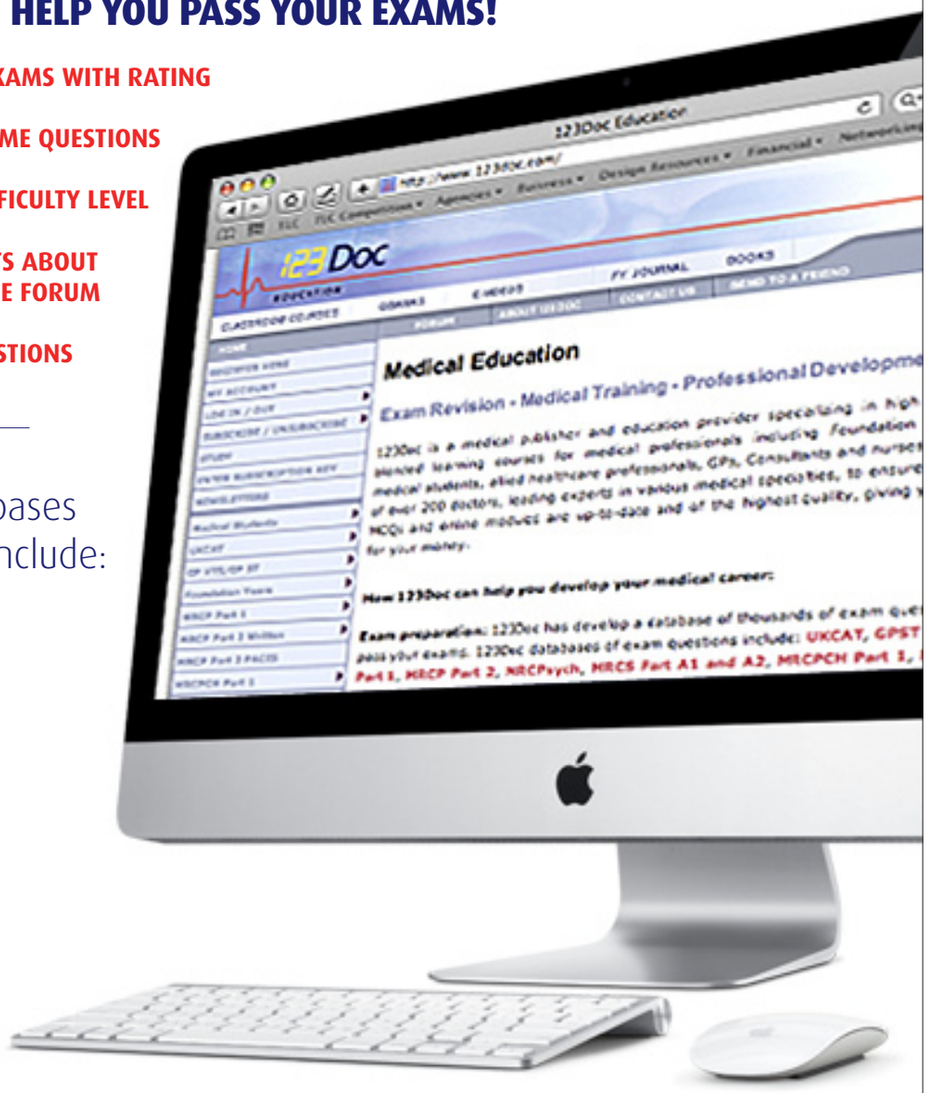

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