



Exam Corner

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The FRCS (Tr & Orth) examination has three components: MCQs, Vivas and Clinical Examination. The Vivas are further divided into five sections comprising Basic Science, Adult Pathology, Hands, Children's Orthopaedics and Trauma. The Clinical Examination section is divided into upper- and lower-limb cases. The aim of this section in the Journal is to focus specifically on the trainees preparing for the exam and to cater to all the sections of the exam every month. The vision is to complete the cycle of all relevant exam topics (as per the syllabus) in four years.

MCQs – Single Best Answer

- 1. Which of the following surgical techniques for spinal deformity carries the highest risk of tetraplegia/paraplegia?**
Answer: a. Anterior thoracic surgery
Anterior thoracic approaches place the segmental blood vessels to the spinal cord at risk. There are various vascular watershed areas in the thoracic spinal cord, including between T4 and T9. This is a particular risk when correcting congenital deformity, particularly kyphosis. Sensory and motor monitoring of the spinal cord is recommended.
Spinal osteotomy also carries a high risk of spinal cord injury. This is particularly seen in correction of kyphosis and is due to stretching of the spinal cord during the correction and a frequent reduction in the cross-sectional area of the spinal canal. Spinal osteotomy in ankylosing spondylitis has been associated with permanent neurological deficit in up to 7.8% of patients.¹
- 2. Which of the following statements is true when comparing a non-anatomic (reattachment of the brachialis muscle) with an anatomic reinsertion of the distal biceps brachii tendon following rupture?**
Answer: e. Decreased strength of supination
Klonz et al² showed that in half of the patients who had undergone brachialis tenodesis, supination strength was only 42% to 56% of the uninjured arm, compared with 91% supination strength following anatomical repair. Flexion strength was 97% following anatomical repair and 96% following brachialis tenodesis.
- 3. A 25-year-old male sustained a traumatic anterior dislocation of the shoulder joint four weeks ago while playing rugby. This is his third dislocation in the last six months. Besides an anteroposterior, axillary and lateral view, what other radiological view will aid your investigation of his shoulder?**
Answer: d. Westpoint view
This view is good for assessing glenoid bone loss. Other useful views include the Stryker view, which will highlight a Hill-Sachs lesion.
- 4. All of the following therapeutic agents used in the management of metastatic disease of bone act by inhibiting the proliferation of tumour cells except:**
Answer: d. Pamidronate
Pamidronate inhibits bone resorption. It adsorbs to hydroxyapatite crystals in bone and may directly block dissolution of this mineral component of bone. Pamidronate inhibits farnesyl pyrophosphate (FPP) synthase, inhibiting osteoclast activity, in turn reducing bone resorption and bone turnover.
- 5. A 40-year-old man presents to an arthroplasty clinic with a painful right hip. A plain radiograph reveals a dysplastic acetabulum with an 80% subluxed femoral head. How should the dysplasia be classified?**
Answer: c. Crowe Grade 3
Gaston et al³ suggested a new classification system for the adult dysplastic hip requiring total hip arthroplasty. This separately grades the femur (F) and the acetabulum (A). The Crowe classification⁴ is one of the most commonly used systems in reconstruction of the adult dysplastic hip. It is based on the magnitude of proximal femoral migration relative to the acetabulum as measured on an AP radiograph of the pelvis:

 - I < 50% subluxation or proximal displacement <10% of pelvic height
 - II 50% to 75% subluxation or 10% to 15% displacement
 - III 75% to 100% subluxation or 15% to 20% displacement
 - IV > 100% subluxation or displacement > 20% of pelvic height
- 6. In fractures of the clavicle treated non-operatively, the risk of nonunion is NOT increased by:**
Answer: b. Male gender
Robinson et al⁵ found that following a diaphyseal clavicle fracture, the risk of nonunion was significantly increased by advancing age, female gender, displacement of the fracture, and the presence of comminution. The risk of nonunion was also higher in lateral and medial clavicle fractures.

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Rotation

Vivas

Adult Pathology

1. Describe the radiograph of this 75-year-old man with a long history of a short leg and stiff hip (Fig. 1).



Fig. 1

Answer: This is an AP radiograph of the left hip and proximal femur. There is gross deformity of the proximal femur with bowing, expansion of the bone, thin cortices and cystic / lytic lucencies. This could be described as a shepherd's crook deformity. The cysts have a 'ground glass' appearance with a well defined sclerotic rim. The femoral head is aspherical and the hip joint is arthritic, with joint space narrowing and subchondral sclerosis. The trabeculae of the femoral head, neck and the periacetabular bones are coarse and expanded.

2. What is the diagnosis?

Answer: This appearance suggests a diagnosis of fibrous dysplasia. It is affecting more than one bone therefore this is polyostotic fibrous dysplasia.

Fibrous dysplasia accounts for approximately 7% of all benign bone tumours. Any bone may be affected. The proximal femur is most commonly affected, followed by ribs, maxilla, tibia, humerus and pelvis.

Due to the coarse appearance of the trabeculae the differential diagnosis would include Paget's disease.

3. Is this an inherited condition?

Answer: No. Most cases of fibrous dysplasia display no particular pattern of inheritance.

4. What is the aetiology?

Answer: It is caused by a genetic mutation of GS alpha protein (chromosome 20q13). This affects the cAMP signalling pathway, leading to increased production of cAMP. There is a failure of production of normal lamellar bone.

5. Discuss the treatment options for this patient.

Answer: This patient has presented with a short leg and a stiff hip. The short leg is most likely due to the proximal femoral deformity. He has an arthritic hip which will probably be causing stiffness. The greater trochanter may be abutting the pelvis upon hip movement, which would also lead to restricted movements. The mechanics of the hip, abductors and proximal femur will undoubtedly be abnormal, leading to further problems when mobilising.

The presence of pain would alert one to the possibility of a pathological fracture. However pain is not mentioned in this case.

The proximal femur is subject to large forces and the patient is therefore at risk of sustaining a pathological fracture (the

bone is dysplastic and weakened and is of an abnormal shape). I would explain this to the patient and counsel him regarding treatment options.

Treatment options would include:

- Regular clinical review and radiographs to monitor for an impending fracture. Malignant transformation is infrequent and has been reported in 0.4% to 4% of cases.
- Simple measures to increase leg length such as a built-up shoe.
- Use of walking aids.
- Bisphosphonate therapy is effective in reducing bone turnover and may reduce the chance of fracture.
- Prophylactic fixation would be difficult or impossible due to the abnormal geometry of the proximal femur.
- Reconstructive surgery:
 - This would be complex and have a high risk of complications for the patient. Dysplastic bone is more vascular than normal bone.
 - Osteotomy and fixation of the femur may not be possible and would not treat the arthritic hip joint. The osteotomy sites will heal with dysplastic bone.
 - Total hip replacement would be difficult due to the abnormal shape of the proximal femur and position of the greater trochanter. This would require an osteotomy and meticulous pre-operative planning. The musculature may also be abnormal and shortened, therefore requiring release.
 - Proximal femoral or total femoral replacement would also be an option.

Trauma

A 26-year-old man was involved in a road traffic accident. He was driving the car and had his seat belt on. This is the radiograph obtained in A&E (Fig. 2).



Fig. 2

1. Describe the radiograph.

Answer: This is a lateral radiograph of the cervical spine. There is anterior translation at the C5/C6 level of around 25% of the width of the vertebral body. There is anterior soft-tissue swelling and blunting of the anterosuperior C6 vertebral body. The C5/C6 facet joint(s) are incongruent and subluxed or dislocated. This is suggestive of a hemi-facet dislocation.

2. What is the mechanism of these injuries?

Answer: According to Allen's mechanistic classification of sub-axial cervical spine injuries this is a distractive flexion (DF) injury, most probably DF stage 2. This is caused by primarily flexion injury vectors that rotate about an axis anterior to the vertebral body.

3. How would you manage this patient?

Answer: I would manage the patient according to ATLS guidelines:

- Supine on a spinal board with triple cervical spine immobilisation (rigid collar, blocks and tape), bed rest and log rolling.
- After primary survey, carrying out any emergent treatment and stabilisation of the patient I would perform the secondary survey, including a full neurological examination, log roll, rectal examination and catheterisation noting the presence / absence of catheter tug sensation and reflex.
- Record any neurological deficit.
- CT scan of the head, whole spine, chest, abdomen and pelvis.
- Transfer the patient to a spinal mattress.
- Pre-operative MRI scan of the cervical spine.
- Serial neurological examination.

Non-operative management

Unilateral facet dislocations in patients without any sign of neurological injury or for those too unwell to undergo surgery can be treated non-operatively in Halo fixation. However, the role of non-operative treatment is minimal.

Operative management

The optimal treatment remains unclear. The principles are to reduce the subluxation / dislocation and then stabilise the spine. There is evidence that facet subluxation / dislocation with neurological compromise should undergo urgent reduction in order to improve the neurological outcome.⁷ This can be achieved either open or closed.

Closed reduction is a safe method in the awake, alert, co-operative patient who can be serially examined.

Pre-reduction MRI to detect a herniated disc is recommended if the patient does not fulfil these criteria or particularly in the incomplete spinal cord injured patient.

A post-reduction MRI scan is usually obtained prior to operative stabilisation to look for a herniated disc.

In a patient who has undergone successful closed reduction, the spine can be stabilised operatively from either anterior or posterior. In the presence of a herniated disc post-reduction, the surgeon may wish to perform an anterior approach to remove the herniated disc. Instrumentation can be performed either anteriorly, posteriorly or using a combined approach for more unstable injuries.

If a closed reduction cannot be achieved, an anterior or posterior approach may aid in reducing a locked facet dislocation. This is usually performed through a posterior approach.

4. What is the role of the MRI scan in the management of these injuries?

Answer: The role of pre-reduction MRI for facet dislocations is controversial. There is potential for neurological deterioration with closed or open reduction.

Disc material may be extruded during the initial injury. Some believe that this material may be pulled back into the spinal cord during reduction, contributing to cord injury. Therefore it has been suggested that MRI should be performed to detect disc herniation. If the disc is herniated, advocates of this technique would proceed to cervical discectomy prior to open or closed reduction.

However, in the awake, alert and examinable patient pre-reduction MRI may not be necessary. Reports suggest that closed reduction can be safely performed, provided that serial neurological examination is possible during this process. Several studies highlight the presence of herniated disc material after successful closed reduction and in the absence of neurological compromise.

Post-reduction MRI can be performed after emergent reduction and will show any discoligamentous disruption facilitating planning for further stabilisation surgery. MRI should be performed urgently if there is neurological deterioration during the reduction procedure.

Hands

A 30-year-old secretary presents with a history of pain over her wrist following a fall (Fig. 3). Radiographs show an incidental finding, and no other bony injury is noted.



Fig. 3

1. What is the diagnosis?

Answer: Carpal synostosis / coalition or lunate-triquetral coalition.

2. What is the incidence of this condition?

Answer: The incidence of carpal synostosis in the white population is 0.2%. It is much more common in black populations, especially West-Africa where this increases to 9.5%. It is often bilateral and some cases may be hereditary. Lunate-triquetral coalition is the most common carpal synostosis, followed by capitate-hamate coalition.

3. What syndromes are associated with this condition?

Answer: Carpal fusions may be associated with both tarsal fusions and with upper limb failure of formation deficiencies:

- Acro-pector-vertebral dysplasia.
- Arthrogyrosis multiplex congenita.
- Nievergelt's syndrome.
- Oto-palato-digital syndrome.
- Bird-headed dwarfism.
- Hand-foot-uterus syndrome.
- Fetal alcohol syndrome.
- Diastrophic dwarfism.
- Turner's syndrome.

4. How is this condition classified?

Answer: Lunate-triquetral coalition is classified according to Minaar's system:

- Proximal pseudarthrosis.
 - Proximal osseous bridge with distal notch.
 - Complete fusion.
 - Fusion with other associated carpal anomalies.
- This patient has a type 2 coalition.

5. How would you manage this patient?

Answer: I would take a history and examine the patient, particularly looking for the area of maximal tenderness. I would then order further imaging accordingly (e.g. scaphoid views if tender in the anatomical snuff-box, clenched fist views if suspecting scapholunate dissociation). Most surgeons believe that congenital carpal synostoses are

asymptomatic and result in no wrist dysfunction. Diagnosis is usually incidental and treatment is unnecessary. There are reports of symptomatic type 1 coalitions that have been treated successfully with operative fusion. With this in mind, I would treat the patient symptomatically (cast or splint, analgesia) and review them in 10 to 14 days with further radiographs and clinical examination. If they remained symptomatic in the presence of normal radiographs (no fracture), I would request further imaging such as a nuclear bone scan or MRI to rule out an occult fracture. If the bone scan was negative and in the presence of continuing symptoms I would request an MRI to rule out a soft-tissue injury.

Children's Orthopaedics

Clinical photograph and selected radiographs (Fig. 4) of a two-year-old child who is alert, able to sit but not standing independently.

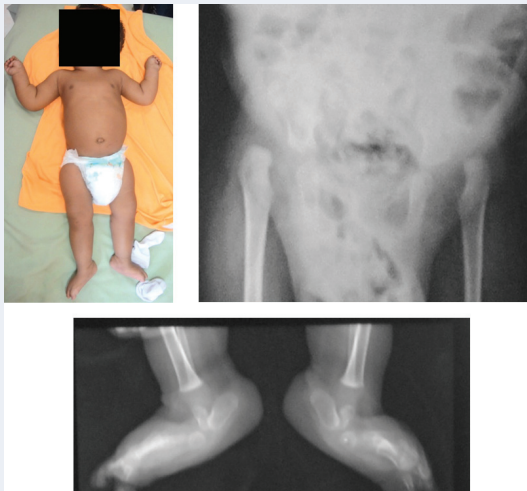


Fig. 4

1. What is the likely diagnosis and how would you manage the case?

Answer: The AP radiograph of the pelvis shows bilaterally dislocated hip joints with proximal migration of the femora. The lateral radiographs of both feet and ankles show bilateral congenital vertical talus (CVT). Clinically the child has bilateral 'rocker bottom feet' that are seen with CVT. The thumbs are adducted and flexed - this is possibly a thumb-in-palm deformity.

These conditions are all associated with a diagnosis of arthrogryposis. It is important to diagnose the exact type of arthrogryposis or arthrogryptic-like syndrome.

Management:

The thumbs are likely to respond well to physiotherapy and splintage but release of any remaining contracture may be indicated after a functional assessment. Physiotherapy is also indicated for the general improvement of mobility.

It is reasonable to correct the feet surgically, followed by prolonged splintage.

However, operative reconstruction of the hips is not indicated. The dislocations are high, the hips are poorly developed and likely to be stiff and the child's walking potential is uncertain.

Clinical photograph (Fig. 5a) and radiograph (Fig. 5b) of a three-year-old child with a progressive deformity of the left tibia.



Fig. 5a

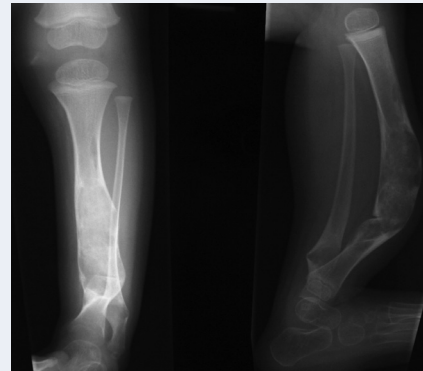


Fig. 5b

2. What is the likely diagnosis, what may be an associated condition and what are the options for treatment?
Answer: The diagnosis is osteofibrous dysplasia, which may be associated with adamantinoma. Treatment is controversial. Prolonged splintage is indicated initially and some cases may resolve spontaneously. Corrective osteotomy and grafting may be indicated where the affected area is small. In this case the lesion is extensive and the deformity is progressive and disabling. Although challenging, excision of the affected area and bone transport should be considered.

Basic Science

1. What are the features of renal osteodystrophy?
Answer: Renal osteodystrophy is the name given to describe the constellation of pathologic bony entities that can occur in the patient with underlying renal disease.

Clinical features:

- Short stature.
- Developmental delay.
- Delay in appearance of secondary growth characteristics.
- All of the clinical findings associated with rickets.
 - Frontal bossing.
 - Rachitic rosary.
 - Increased thoracic kyphosis - 'rachitic catback'.
 - Generalised shortening of long bones and widening of joints.
 - Varus humeri.
 - Genu varus or valgus.
 - Ligamentous laxity, with angular deformities exacerbated by weight-bearing.
- Infections and pathological fractures - frequently secondary to steroid treatment / dialysis.
- Bone tenderness.
- Soft-tissue itching and irritation secondary to ectopic calcification.
- Joint pain and decreased range of movement secondary to ectopic calcification.
- Gait disturbances.
- Radiographic findings.
 - "Salt and pepper skull"

- Loss of the cortical outline of the distal 1 cm of the clavicle.
- Subperiosteal bone resorption – ulnae, phalangeal tufts, medial proximal tibiae.
- Brown tumours – lytic lesions often in the pelvis or long bones.
- Slipped epiphyses. Due to hyperparathyroidism caused metaphyseal bone resorption and epiphyseal lysis.
 - Most commonly proximal femur but can be proximal humerus, distal femur or distal tibia.
 - Distal radius and ulna occurs in older children.

2. What is the pathophysiology for the development of renal osteodystrophy?

Answer: Pathophysiology:

- Damage to the phosphate tubules of the kidney leads to phosphate retention and reduced production of 1,25-dihydroxy vitamin D.
- Lack of 1,25-dihydroxy vitamin D leads to an inability of the gut to absorb calcium.
- Hypocalcaemia leads to secondary hyperparathyroidism and bone resorption.

3. What are the effects of renal osteodystrophy?

Answer: There are four clinical entities:

- Rickets or osteomalacia – reduced mineralisation of osteoid.
- Osteitis fibrosa cystica – lytic bone lesions secondary to increased levels of PTH.
- Osteosclerosis – due to increased numbers of bony trabeculae. Most evident in the long bones and spine.
- Ectopic calcification – related to the hyperphosphataemia of renal patients. They are typically acidotic which leads to an increased serum solubility of calcium salts. If serum calcium returns to near normal levels (dialysis or diet related) there will be a precipitation of calcium into areas such as the corneas, conjunctivae, skin, arterial walls and periarticular soft tissues.

4. What is the treatment of patients with renal osteodystrophy?

Answer: Treatment is by a multidisciplinary team including a nephrologist, endocrinologist and orthopaedic surgeon.

- Management of the underlying renal disease – medical therapy, steroids, dialysis, renal transplant.
- Drugs to control calcium and phosphate levels.
- Parathyroidectomy is sometimes required.
- Careful vitamin D replacement.
- Surgical treatment of epiphyseal slips.
- Limb deformity correction.

5. What are phosphate-wasting conditions?

Answer: Phosphate wasting conditions are inherited diseases of renal phosphate handling that lead to urinary phosphate wasting and depletion of total body phosphorus stores.

More than 99% of total body phosphorus stores are located intracellularly therefore serum phosphorus concentration does not accurately reflect total body phosphorus stores. Depending on the age of onset of renal phosphate wasting, patients may present with growth retardation, skeletal

deformities of the upper and lower extremities (genu varum or genu valgum), insufficient weight gain, metaphyseal widening, palpable enlargement of the costochondral junctions (rachitic rosary), and frontal bossing. There are several distinct forms of hereditary renal phosphate wasting:

- X-linked dominant hypophosphataemic rickets (XLH). (Most common form).
- Autosomal dominant hypophosphataemic rickets (ADHR).
- Autosomal recessive hypophosphataemic rickets (ARHR).
- McCune-Albright syndrome (MAS).
- Hereditary hypophosphataemic rickets with hypercalciuria (HHRH).
- Autosomal recessive Fanconi syndrome and hypophosphataemic rickets (ARFS).

6. What is the role of FGF in phosphate-wasting conditions?

Answer: FGF-23 has a central role in the regulation of phosphorus. It inhibits renal phosphate re-absorption and inhibits the synthesis of 1,25-dihydroxy vitamin D₃. FGF-23 acts on the proximal tubule in the kidney, leading to phosphaturia. Various abnormalities of FGF-23 or its associated pathways lead to hereditary renal phosphate wasting (see table below).

Disease	Gene	FGF-23 level	1,25-(OH) ₂ vitamin D ₃ level	Hypercalciuria
XLH	PHEX	Elevated	Normal or low	No
ADHR	FGF-23	Elevated	Low	No
ARHR	DMP1	Elevated	Normal or low	No
ARHR	ENPP1	Normal / elevated	Normal	No
MAS	GNAS	Elevated	Not known	No
HHRH	SLC34A3	Normal or low	Elevated	Yes
ARFS	SLC34A3	Low to normal	Decreased	No

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