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Quiz

- 1) Fibrodysplasia ossificans progressiva is characterized by which one of the following condition?
 - A. Constriction bands in wrist and ankle
 - B. Short valgus great toe
 - C. Blue sclera
 - D. Down-pointing umbilicus
- 2) In trigger finger the level of tendon sheath constriction is found at the level of:
 - A. Middle phalanx
 - B. Proximal interphalangeal joint
 - C. Proximal phalanx
 - D. Metacarpophalangeal joint
- 3) In limb replantation, last to be repaired is:
 - A. Artery
 - B. Vein
 - C. Nerve
 - D. Extensor tendon
- 4) Identify the structure marked with arrow in Fig. 1:
- 5) Schaeffer's test is used for
 - A. Testing DRUJ stability
 - B. Diagnosing fracture of ulnar styloid
 - C. Confirming the presence of Palmaris Longus
 - D. Diagnosis of carpal tunnel syndrome

Answers and explanations

Answer 1): B. Short valgus great toe

Fibrodysplasia ossificans progressiva (FOP) is a very rare autosomal dominant genetic disorder with a prevalence of one case in 2 million individuals. FOP is caused by a recurrent activating mutation in the gene encoding activin receptor 1A/activin-like kinase 2. The name myositis ossificans progressive was assigned to the condition by von Dusch in 1868.

Heterotopic ossification (HO) typically begins to form during the first decade of life preceded by painful soft tissue swellings (flare-ups). These swellings are sometimes mistaken for aggressive fibromatosis or musculoskeletal tumors. These formations of heterotopic bone usually span over joints and lead to ankylosis and progressive immobility. The HO usually follows a specific anatomic pattern. They begin in the neck, shoulders, back and then progress to trunks and limbs.

Malformations of great toes, such as hallux valgus, deformed proximal phalanges and shortened first metatarsal bones are pre-osseous features of FOP. The malformation of

great toes can vary from a fibular deviation of the great toes to their complete absence.

Many patients develop severe scoliosis and thoracic insufficiency. Other spinal abnormalities seen are large posterior elements, tall narrow vertebral bodies and fusion of facet joints in cervical spine. Patients with FOP also have abnormalities in the rest of the skeleton like clinodactyly, short broad femoral necks, and osteochondromas.

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Answer 2): D. Metacarpophalangeal joint

Stenosing tenosynovitis (trigger finger) is usually seen in individuals older than 45 years of age. It can have various causes like rheumatoid, infectious, tumors of synovial sheath, hormonal and mechanical factors. Patients may note a lump or knot in the palm that can be palpated by the examiner's fingertip and moves with the tendon. Local tenderness may be present. Triggering is accentuated by pressure. This nodule is usually at the level of the metacarpophalangeal joint at the entry of the tendon into the proximal annulus. The first annular pulley (A1) is by far the most often affected pulley, though cases of triggering at second and third annular pulleys have also been reported. The lump may be the thickened area in the first annular part of the flexor sheath or a nodule or fusiform swelling of the flexor tendon just distal to it. Pressure accentuates the snapping or triggering of the distal joints.

When associated with a collagen disease, the long and ring fingers are most commonly involved. In rheumatoid arthritis, the nodule might be distal to the metacarpophalangeal joint and is not always relieved by sectioning the A1 pulley alone. Occasionally, a partially lacerated flexor tendon at this level heals with a nodule sufficiently large to cause triggering.

Treatment of trigger finger is usually nonoperative by measures like stretching, night splinting, and combinations of heat and ice. A single corticosteroid injection has a 60% success rate. A percutaneous release using a fine needle can lead to incomplete release and damage to flexor tendons and

Fig. 1 – Clinical photograph of the child's neck from behind.

digital nerves. Surgical release reliably relieves the problem for most patients.

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Answer 3): C. Nerve

Replantation is replacement of tissue or a structure (digit) in the site from which it was previously lost or removed. For replantation; at the trauma scene, collect and preserve all amputated parts. Even parts that are crushed and not thought to be suitable for replantation can provide tendons or bone.

The amputated part must be cooled to 4 °C to preserve it. An hour of warm ischemia is equivalent to approximately 6 h of cold ischemia. Hence, cooling can prolong the window of opportunity for replantation. Wrap the part in saline-soaked gauze or towel and then place it in a dry plastic bag. Place this bag on ice in an icebox/cooler, or in a second bag filled with ice and water, as soon as possible. This 2-layer approach avoids submersion of the part in ice water, which causes freezing of the tissues and cell destruction.

If possible, bleeding from proximal vessels should be controlled with pressure, and ligature control should be avoided. Bone, tendon and skin can tolerate approximately 8–12 h of warm ischemia and as long as 24 h of cold ischemia. However, muscle necrosis after 6 h of warm ischemia or 12 h of cold ischemia. In general, amputated digits may tolerate 12 h of warm ischemia and 24 h of cold ischemia. Other major amputations tolerate 6 h of warm ischemia and 12 h of cold ischemia because of their larger muscle content.

The normal sequence of replantation is as follows:

1. Debridement
2. Identification and/or tagging of vital structures
3. Skeletal stabilization
4. Extensor tenorrhaphy
5. Venous repair
6. Artery repair
7. Repair of flexor tendon
8. Neurorrhaphy
9. Skin closure
10. Dressing

The amount of bone shortening necessary is dictated by the extent of comminution. Shortening to facilitate vascular repairs is not necessary because vein grafting is adequate. Following appropriate shortening, the bone may be stabilized. Following bone and/or joint stabilization, the extensor tendon mechanism is repaired to improve stability. Veins are repaired prior to arterial anastomosis. This approach minimizes blood loss and prevents a bloody surgical field. In situations of prolonged ischemia arteries are repaired after repair of just one vein. Flexor tendons may be repaired prior to or following neurovascular anastomosis. In digital amputations it is preferred to add stability by repairing flexor tendons prior to arterial and digital nerve repair. For more proximal amputations, muscles and tendons are reconnected after performing the arterial repair. The nerve is the last structure to be repaired.

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Answer 4): Omovertеbral bar in Sprengel shoulder

Sprengel deformity, or congenital elevation of scapula, is a congenital deformity of the shoulder named after Otto Gerhard Karl Sprengel (1852–1915), a German surgeon who described four such cases in 1891.

Sprengel deformity is usually noticed at birth, elevated scapula being visually noticeable and is associated with restricted movements of the scapula and shoulder. Failure of caudal migration of the scapula during early fetal development is implicated to result the deformity. It usually coexists with other congenital abnormalities of vertebrae and ribs. Omovertеbral bar (fibrous, cartilaginous and/or osseous connection between scapula and cervical spine) is often present. It is also commonly associated with hypoplasia or atrophy of regional muscles, and these associated features can cause further misshaping of the shoulder and limitation of shoulder movements.

Patients with Sprengel deformity often have other abnormalities like Klippel-Feil syndrome, spina bifida, kyphoscoliosis, torticollis and underdevelopment of clavicle or humerus which should be looked for in any patient presenting with Sprengel deformity.

Cavendish classification is commonly used for grading of this condition. Grade I cases have mild deformity and the deformity is almost invisible when the part is covered with clothes. Grade II deformity is still mild but appears as bump

because of the convex superomedial portion of the high scapula. Grade III is moderate deformity with 2–5 cm of visible elevation of the affected shoulder. Grade IV is severe deformity with >5 cm elevation of the affected shoulder, accompanied by neck webbing. The affected scapula is elevated and rotated, with the inferior angle directed laterally.

The radiographic appearance is graded by Rigault's classification. Grade I involvement is described when superomedial angle of scapula is lower than T2 vertebra but above T4 transverse process. In grade II, the superomedial angle is located between C5 and T2 transverse process and in grade III, the superomedial angle is above C5 transverse process.

Surgery is performed to improve cosmetic and functional disability. It is generally done between 3 and 8 years of age in patients having moderate to severe disability (or a Cavendish score of 3–4). Two of the most used surgical methods are the “Woodward” procedure and the “modified-Green” procedure with good functional and cosmetic outcome.

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Answer 5): C. Confirming the presence of Palmaris Longus

Palmaris Longus, a slender fusiform muscle lying medial to the flexor carpi radialis originates from the medial epicondyle by a common flexor tendon, from the adjacent intermuscular septa and antebrachial fascia. It then passes anteriorly to the flexor retinaculum and is attached to its distal half and centrally to the palmar aponeurosis often sending a tendinous slip to the thenar muscles. PL is a functionally redundant but an accessible muscle.

Its absence appears to be hereditary though genetic transmission is not clear. Understanding its variations is useful because it is often used as tendon graft and for tendon transfer. PL is a phylogenetically degenerated muscle as suggested by a short muscle belly and long tendon as well as the replacement of a distal tendon by the ligamentous palmar aponeurosis. It is a weak flexor of the wrist, and anchors the

Fig. 2 – Standard test (Schaeffer's test) to assess the presence of Palmaris Longus.

Fig. 3 – Thompson's test to assess the presence of Palmaris Longus.

Fig. 4 – Mishra's test I to assess the presence of Palmaris Longus.

skin and fascia of the hand against the shearing forces. PL agenesis varies with race, sex, and to the right and left side. There is a wide variation in the incidence of PL ranging from 0% to 63% with an overall 16% unilateral and 9% bilateral absence described in the literature.

Fig. 5 – Mishra's test II to assess the presence of Palmaris Longus.

Fig. 6 – Pushpakumar's “two-finger sign” method to assess the presence of Palmaris Longus.

Various tests described to detect its presence are:

1. Standard test (Schaeffer's test): The subject is asked to oppose the thumb to the little finger and then flex the wrist (Fig. 2).
2. Thompson's test: The subject is asked to make a fist, then flex the wrist and finally the thumb is opposed and flexed over the fingers (Fig. 3).
3. Mishra's test I: The metacarpophalangeal joints of all fingers are passively hyperextended by the examiner and the subject is asked to actively flex the wrist (Fig. 4).
4. Mishra's test II: The subject is asked to abduct the thumb against resistance with the wrist in slight palmar flexion (Fig. 5).
5. Pushpakumar's “two-finger sign” method: The subject is asked to fully extend the index and middle finger, the wrist and other fingers are flexed and finally the thumb is fully opposed and flexed (Fig. 6).

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Saravpreet Ahluwalia, MS Ortho, MRCS
Senior Resident, Department of Orthopaedic Surgery, Maulana Azad
Medical College Associated Lok Nayak Hospital,
New Delhi 110002, India

Nishit Bhatnagar, MBBS
Post Graduate Student, Department of Orthopaedic Surgery,
Maulana Azad Medical College & Associated Lok Nayak Hospital,
New Delhi 110002, India

Sumit Arora, MS Ortho, DNB Ortho, MNAMS*
Assistant Professor, Department of Orthopaedic Surgery, Maulana
Azad Medical College & Associated Lok Nayak Hospital,
New Delhi 110002, India

*Corresponding author. c/o Mr. Sham Khanna, 2/2, Vijay Nagar,
Delhi 110009, India. Tel.: +91 9868329389.
E-mail address: mamc_309@yahoo.co.in
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