



# POSNA

## The Core Curriculum

### **Congenital deficiencies of the upper limb**

#### **Objectives**

1. Describe the embryology and formation of the upper limb bud
2. Describe the classification for upper limb deficiencies adopted by the American Society for Surgery of the Hand and the International Federation of Societies for Surgery of the Hand
3. Discuss the timing of prosthetic fitting for failure of transverse formation
4. Discuss strategies for management of failure of longitudinal formation
5. Discuss strategies for management of failure of differentiation
6. Define and discuss strategies for management of clinodactyly and camptodactyly
7. Discuss the pathologic anatomy and strategies for management of delta phalanx
8. Discuss the genetic transmission of ulnar polydactyly
9. Define Madelung's deformity
10. Describe the natural history of congenital trigger thumb

#### **Discussion**

Development of the limb bud is discussed under embryology. The upper limb bud is derived from the lateral plate scleroderm, with the apical ectodermal ridge first evident at the 4th week. Under the regulation of elaborate genetic control, especially the homeobox genes, limb formation is complete in the 8th week. The classification system developed by Swanson is now internationally accepted, and divides congenital anomalies into seven categories, failure of formation (transverse or longitudinal), failure of differentiation, duplication, overgrowth, undergrowth, constriction bands, and generalized skeletal abnormalities. A number of anomalies of the upper limb are associated with cardiac anomalies, hematologic problems, and VATER syndrome. Poland's syndrome includes hypoplasia of the pectoralis major.

Failure of transverse formation is a congenital amputation, described by the affected region (midarm, proximal forearm, wrist, midhand, etc). The proximal forearm is most common. A passive prosthetic device is fitted by about 6 months to allow the child to incorporate the limb in development. Longitudinal deficiencies can involve either deficiencies of the radial or ulnar paraxial segments in varying degrees of severity. The radial deficiency, or radial club hand is more common, The thumb is usually deformed or absent. The most severe variety is most common. Treatment strategies include stretching and splinting for mild deformities and centralization of the ulna (usually by 1 year of age) followed by pollicization of the thumb for severe deficiencies. Ulnar

clubhand is less frequent, and has less severe functional deficiencies so it is usually managed conservatively. Central deficiencies are known better as lobster claw hands, which can rarely be improved functionally. Failure of differentiation is manifest clinically as syndactyly, which is quite common, 1:2000 live births. Surgery is generally planned for the first or second years of life depending on the complexity. Camptodactyly is a flexion deformity of the PIP joint, usually the little finger. Although surgical procedures are available, it is unusual that surgery results in meaningful functional improvement. Clinodactyly is lateral deviation of a digit. It is usually bilateral, usually involves the little finger, and is transmitted as an autosomal dominant trait. Treatment is seldom indicated. Delta phalanx is well described by the name; the epiphysis is triangular and the finger distal to the delta phalanx is angulated. This condition is sometimes known as a longitudinal epiphyseal bracket, as the physis is bracketed between the two ends and tethered on the shorter side. Treatment in the hand has been confined to osteotomy; Mubarak has described division of the bracket in the great toe. Congenital trigger thumb is fairly common. About 30% of trigger fingers detected at birth will resolve spontaneously, so surgery should be deferred at least until after age 1. Surgery is identical to that performed for trigger finger at any age. Duplication of any ray in the hand is possible, it is most common on the ulnar side; the incidence of this duplication is 1:300 in the black population. Treatment varies in difficulty depending on the amount of the ray duplicated. Madelung's deformity is characterized by ulnar and volar tilt of the distal radial epiphysis, with an early fusion of the distal ulnar physis. Similar abnormalities can be seen with a number of the chondroplasias and Ollier's disease.

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