

## CME Article

# Common paediatric orthopaedic problems in the upper limb

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In this article, we attempt to provide guidance for conducting the paediatric examination in an orderly, comprehensive sequence to arrive at the diagnosis in common musculoskeletal problems

of the upper limb in children and adolescents, paying particular attention to clinical problems such as cubitus varus, polydactyly and Sprengel's shoulder.

## CONGENITAL MUSCULAR TORTICOLLIS

### Description

Torticollis or wry neck is defined as a condition where the head is tilted toward one side and the chin is pointing in the opposite direction. Congenital muscular torticollis (CMT) is a postural deformity detected at birth or shortly after birth.

### Frequency

CMT has an estimated incidence of 1-2% and male-to-female ratio is 3:2 in the Chinese population<sup>(1)</sup>.

### Location

75% of CMT cases involve the right side.

### Pathogenesis

CMT (present at birth) may be caused by malpositioning of the head in the uterus, or by prenatal injury to the sternocleidomastoid muscle (SCM) that results in fibrosis or soft tissue compression leading to compartment syndrome, as evidenced by CMT being more frequent after breech presentations and with congenital hip dysplasia<sup>(2)</sup>. In rare instances, bony anomalies of the cervical spine include the Klippel-Feil syndrome, or ocular imbalance might be causative. Other possibilities are spontaneous subluxation of a cervical vertebra, cervical adenitis secondary to upper respiratory tract infection, unilateral soft tissue infection, neck tumours or myositis.

### Diagnosis

Most cases can be diagnosed after history and physical examination. Physical examination should include careful examination of the head and neck. Possible associated conditions include developmental dysplasia of the hip and clubfoot, and careful neurological examination must be done. Radiographs of the cervical spine should be obtained to rule out obvious bony abnormality.

### Significance

CMT causes plagiocephaly (a craniofacial deformity), facial asymmetry (hemihypoplasia) (Fig. 1), scoliosis and atrophy of the ipsilateral trapezius muscle, if not corrected.

### Treatment

Treatment of CMT involves manual stretching of the shortened neck muscle. Passive stretching and positioning are treatments used in infants and small children. Botulinum toxin (Botox) could enhance the effectiveness of stretching on the side of the contracture and allow strengthening of overstretched and weakened muscles on the opposite side of the neck. Surgical release of the SCM is indicated if symptoms persist after one year of age despite conservative treatment, or for children who entered conservative treatment at older ages whereby lateral bending is still  $>15^\circ$  or facial hemihypoplasia has developed. Surgical techniques to lengthen tight SCMs include unipolar release or bipolar release. The technique employed depends upon the severity. Postoperative physical therapy consisting of range-of-motion exercises is recommended after surgical release of the SCM.



**Fig. 1** Photograph of a 3-year-old girl with left torticollis. Note the hemihypoplasia.

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## SPRENGEL'S SHOULDER

### Description

Sprengel's shoulder is a condition where there is a congenital elevation of the scapula. Developmentally, the scapula has failed to descend from its embryological position in the neck.

### Frequency

The male-to-female ratio is 3:1.

### Location

The left side is more commonly affected than the right side.

### Pathogenesis

The aetiology of Sprengel's shoulder is related to limb bud formation. It is due to failure of descent of the scapula during embryonic development from its position in the neck to its normal position in the posterior thorax. The omovertebral bone (a structure of cartilage or bone), connecting the superomedial scapula to the posterior elements of the cervical vertebrae, has a major role in determining the shape and the malpositioning of the scapula<sup>(3)</sup>.

### Diagnosis

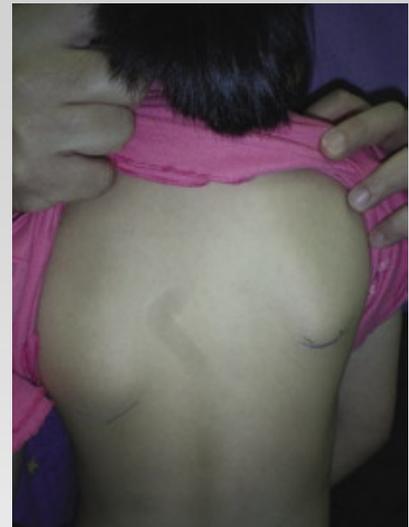
The physical examination usually reveals scapular hypoplasia and asymmetry (Fig. 2), and the presence of an omovertebral bone restricts scapular movements, and therefore abduction of the arm. Radiographs to determine the severity of the Sprengel's shoulder are necessary, as well as to rule out any neck or spinal abnormalities. Radiographs will demonstrate an omovertebral bone in 30%-40% of cases<sup>(4)</sup>.

### Significance

The omovertebral bone is seen in 30% of patients with Sprengel's shoulder. Other congenital malformations are either absent, or include fused ribs, chest wall asymmetry, Klippel-Feil syndrome, cervical ribs, congenital scoliosis, and cervical spina bifida.

### Treatment

Non-operative management consists of physical therapy to maintain the range of motion preoperatively, and segmental strengthening of the muscles in the scapular region. Surgery is best performed when the child is between three and eight years of age and for whom the deformity is marked, or the restriction of motion is severe. The surgery usually consists of release of the tight band, removal of the omohyoid bone that tethers the scapula in its abnormal position, and transplantation of the muscular origins of the scapula to a lower level to keep the scapula in a more normal position.



**Fig. 2** Photograph of right Sprengel's shoulder. Note the asymmetry of the scapulae.

## CUBITUS VARUS

### Description

Cubitus varus is a deformity of the elbow, due to lateral angulation of the joint, and is accompanied by deviation of the forearm towards the midline of the body when the forearm is extended. It is also called gun stock deformity.

### Frequency

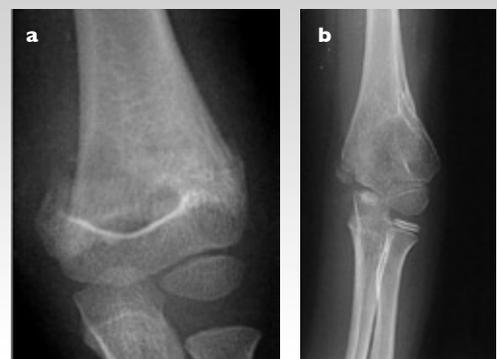
Range from 5% of those fractures treated percutaneously and in 33% of patients treated with traction<sup>(5)</sup> (Fig. 3).

### Location

Both sides are equally affected.

### Pathogenesis

Cubitus varus (Fig. 4) is the most common long-term complication resulting from a malunion of a supracondylar fracture of the humerus in children. The distal humerus remodels in the plane of motion, but significant remodeling in the coronal plane does not occur. Cubitus varus resulting from inadequate reduction or failure to maintain reduction has long been considered a strictly cosmetic problem.



**Fig. 3** Frontal radiographs show (a) poorly-treated fracture of the supracondylar humerus that has subsequently resulted in (b) cubitus varus deformity.

### Diagnosis

The normal carrying angle (i.e. the angle between the upper arm and forearm, normally with a slight valgus deviation) in males ranges from 5° to 10° and in females from 10° to 15°. The cubitus varus deformity is best assessed by comparing the injured elbow with the contralateral side, and is most obvious when the arms are elevated and the elbow is extended (Fig. 4). If the carrying angle is less than 5°, cubitus varus is identified (Fig. 5). This analysis is aided by the use of the Baumann angle (evaluated on the anteroposterior radiograph), which is the angle between the long axis of the humeral shaft and the growth plate of the capitellum. The normal angle is about 85° to 89°. When compared with the opposite uninjured side, Baumann's angle should not be more than 5° different. An increase in Baumann's angle indicates a varus alignment (cubitus varus).

### Significance

Cubitus varus deformity (Fig. 5) is primarily a cosmetic deformity and does not affect function. It may cause other delayed consequences including increased risk of late lateral condyle fracture and posterolateral rotary instability of the elbow.

### Treatment

Corrective supracondylar osteotomy is performed not only for cosmetic appearance, but also to avoid tardy posterolateral rotatory instability of the elbow in the future<sup>(6)</sup>.



**Fig. 4** Left cubitus varus. Photograph shows the position in which cubitus varus is most evident, i.e. arms elevated and elbows extended.



**Fig. 5** Photograph shows the carrying angle with a varus deviation in the left arm, indicating left cubitus varus.

## CONGENITAL RADIOULNAR SYNOSTOSIS

### Description

Congenital radioulnar synostosis is a condition where there is fusion of the superior radioulnar joint.

### Frequency

Males and females are equally affected. About 40% of the cases are unilateral, and 60% are bilateral.

### Location

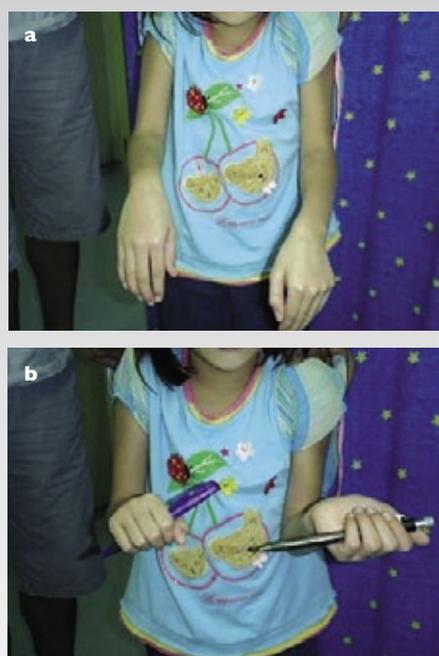
The proximal one-third of the forearm is the most common site of involvement.

### Pathogenesis

Radioulnar synostosis is one of the more common failures of separation of parts of the upper limb. While most cases of congenital radioulnar synostosis occur spontaneously, approximately one in five cases are associated with a family history of the same condition. Congenital radioulnar synostosis can also occur as part of an underlying syndrome or with other developmental abnormalities.

### Diagnosis

The condition is detected only at the age of three or four years when the child uses the hand for various activities. Congenital radioulnar synostosis is diagnosed by treating physicians after



**Fig. 6** Right radioulnar synostosis. Photographs show that from (a) a bilaterally-pronated position, (b) the patient is unable to supinate the right forearm.

a thorough medical history and careful physical examination. There is inability to supinate the forearm as the fusion is mostly in the fully pronated position (Fig. 6). Children initially may have reduced radial heads and later develop symptomatic radial head subluxation as adolescents. Radiographs or computed tomography may be used to confirm the diagnosis.

### Significance

Although most cases have been isolated, additional anomalies may appear, such as clubfoot, developmental dysplasia of the hip (DDH), knee anomalies, hypoplasia of the thumb, polydactyly, syndactyly, carpal coalition, symphalangism and Madelung's deformity.

### Treatment

In general, treatment recommendations for congenital radioulnar synostosis are made based on the degree of functional loss associated with loss of forearm rotation. Patients with unilateral or bilateral deformities with less than 30° of pronation generally do not require surgery. Surgery is needed when severe pronation deformities (more than 60°) cause significant functional difficulty, especially with activities requiring supination. Various surgical options have been proposed to restore the correct position and mobility. These include resection of the synostosis, reconstruction with interpositional materials, rotational osteotomy through or distal to the synostosis mass and recently, success has been reported following vascularised fascio-fatty graft<sup>(7)</sup>.

## RADIAL CLUB HAND

### Description

Radial club hand is a type of congenital hemimelia characterised by a partial or total absence of the radius and the thumb.

### Frequency

Incidence of radial club hand varies between one per 55,000 and one per 100,000 live births.

### Location

Radial club hand is bilateral in 50% of cases.

### Pathogenesis

Radial deficiencies are categorised as a longitudinal failure of formation<sup>(8)</sup>. Muscle deficiencies – those originating on the postaxial side, i.e. the common flexor origin, are more common while those arising from the preaxial side, i.e. the common extensor origin, are rare and tend to be associated with other abnormalities. Skeletal malformations occur not only in the radius but also in the whole upper limb. It is usually associated with an absent or underdeveloped thumb.

### Diagnosis

Careful clinical examination is used to assess the degree of involvement. The infant holds the wrist palmarflexed and radially deviated. The shoulder, elbow, wrist, and digital range of movement are evaluated for active and passive motion. The clinical presentation can be mild (hypoplasia of radius with affected thumb); moderate (deficient thumb and partial absence of the radius) (Fig. 7); or severe (absence of thumb and entire radius). The thumb is examined for hypoplasia and other anomalies of the fingers usually coexist. The radiographs can be useful in assessing the degree and course of the aplasia, and laboratory analysis includes tests for anaemia and thrombocytopenia.

### Significance

Radial club hand has been associated with many congenital syndromes including Holt-Oram syndrome (cardiac septal defects), thrombocytopenia with absent radius (TAR), Fanconi anaemia (aplastic anaemia), and vertebral, anal, tracheal, cardiac, oesophageal, renal, and limb abnormalities (VACTER syndrome). If the radial club hand is associated with a syndrome, the prognosis depends on it.



**Fig. 7** Moderate right radial club hand. Photographs (a,b) show absence of the thumb and partial absence of the radius.

### Treatment

Correction of radial club hand requires a combination of non-operative and operative management that begins shortly after birth. The primary treatment consists of passive stretching followed by serial casts which are used to stretch tight structures on the radial side in order to achieve longitudinal alignment. When the soft tissues are stretched out, the carpus is surgically centralised on the lower end of the ulna, with the wrist in the corrected position. This procedure is performed in patients aged approximately one year. The pollicisation of the index finger (i.e. reconstruction of the index into a thumb) can be done later to obtain a functioning hand.

## POLYDACTYLY AND SYNDACTYLY

### Description

Polydactyly is the occurrence of extra fingers or toes, while syndactyly is the incomplete separation of fingers or toes, sometimes called webbed fingers or toes.

### Frequency

The prevalence of polydactyly with or without an associated malformation varies between 5 and 17 per 10,000 live births. Syndactyly occurs in 2 to 3 per 10,000 live births, bilateral involvement is found in 50% of patients, and the male-to-female ratio is 2:1.

### Location

In polydactyly, the little finger is the finger that is most often affected, and syndactyly is most common between the middle and ring fingers.

### Pathogenesis

Polydactyly and syndactyly can both occur as isolated conditions or in conjunction with other symptoms as one aspect of a multi-symptom disease. Polydactyly may occur in families as a dominantly inherited trait. The extra digits may be made of only skin and flesh, or skin, flesh and bone. There are many sub-classifications of this anomaly but the two major subgroups are preaxial and postaxial. Pre-axial polydactyly occurs on the side of the thumb or big toe (Fig. 8). Postaxial polydactyly is relatively more common and occurs on the side of the little finger or lateral side of the foot and in association with trisomy 13.

Syndactyly occurs when normal separation of the fingers does not occur by preprogrammed cell death that normally occurs. Syndactyly can be an isolated finding, or it can be found in association with other abnormalities, such as polydactyly, cleft hands, ring constrictions, or craniofacial syndromes (e.g. Apert syndrome)<sup>(9)</sup>. There are two types of syndactyly: simple syndactyly – which involves fusion of only tissues of the fingers, and complex syndactyly – which involves fusion of the bones. A severe form is lobster claw or split hand/foot syndrome (ectrodactyly).

### Diagnosis

Polydactyly and syndactyly can be diagnosed by clinical examination, radiographs, and foetal ultrasonography.

### Significance

The prognosis for isolated polydactyly and syndactyly is excellent. When polydactyly or syndactyly are part of a larger condition, the prognosis depends on the condition.

### Treatment

The goals of surgery are to improve the appearance of the hand and to prevent progressive deformity from developing as the child grows. Polydactyly can be corrected by surgical removal of the extra digit or partial digit. If the main finger has functioning tendons and nerves, the extra digit may be excised to improve the appearance of the hand. The main treatment of syndactyly is surgery to separate the joined parts and more importantly, to add skin grafting on the raw areas. The more fingers involved and the more complex the syndactyly, the earlier the release should be performed.



**Fig. 8** Photograph shows polydactyly of the thumb.

## CONGENITAL TRIGGER THUMB

### Description

Trigger thumb is a condition where the thumb is locked in the flexed position at the interphalangeal joint (IP joint) (Fig. 9). The condition may present at birth, or during infancy.

### Frequency

One-third of patients with congenital trigger thumb have a positive family history of trigger thumbs, and the incidence is reported in less than 0.3% in childhood<sup>(10)</sup>.

### Location

20%-30% of cases are bilateral.

### Pathogenesis

Trigger thumb is due to thickening and constriction of the fibrous flexor sheath that surrounds the flexor pollicis tendon at the base of the thumb. The constriction does not allow for free movement of the tendon within the sheath. A nodular thickening of the tendon may be felt at the base of the metacarpophalangeal joint (Notta's nodule).

### Diagnosis

The condition may present at birth, or during infancy, and is usually not noticeable until infants begin to use their hands. Trigger thumbs are diagnosed by the treating physician after a thorough medical history and careful physical examination. Children with trigger thumbs typically present with a flexed thumb. Early on, the thumb can be extended. However, in the later stages, the thumb will remain in a flexed position. Often, a palpable nodule is felt at the base of the thumb in the palm. No special tests or radiographs are required.

### Significance

If the condition is allowed to progress, the nodule may swell to the point where it gets caught and the finger is locked in a flexed position.

### Treatment

Treatment is often conservative. About 30% of trigger fingers detected at birth will resolve spontaneously, so surgery should be deferred to at least until after the age of one<sup>(11)</sup>. Parents are taught to stretch the thumb and gently extend the thumb, even in the thumb that is locked in flexion. Over a period of 18 to 24 months, the triggering may resolve spontaneously. In cases where the thumb is locked in acute flexion and does not allow any extension, surgical release of the fibrous flexor sheath is indicated by the age of two years to prevent permanent contracture. Surgery involves releasing the A1 pulley overlying the flexor tendon. Care must be taken to avoid the radial digital nerve to the thumb which crosses the metacarpal head at the level of the pulley.



**Fig. 9** Photograph shows congenital trigger thumb in a one-year-old child.

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## SINGAPORE MEDICAL COUNCIL CATEGORY 3B CME PROGRAMME

### Multiple Choice Questions (Code SMJ 200608A)

	True	False
<b>Question 1:</b> The following statement(s) regarding that of radial club hand is/are true:		
(a) Radial deficiencies are categorised as a longitudinal failure of formation.	<input type="checkbox"/>	<input type="checkbox"/>
(b) Muscles arising from the postaxial side are commonly affected.	<input type="checkbox"/>	<input type="checkbox"/>
(c) It is usually associated with an absent or under-developed thumb.	<input type="checkbox"/>	<input type="checkbox"/>
(d) The primary treatment of radial club hand is surgical.	<input type="checkbox"/>	<input type="checkbox"/>
 <b>Question 2:</b> Which of the following is/are risk factor(s) for torticollis?		
(a) Developmental dysplasia of the hip (DDH).	<input type="checkbox"/>	<input type="checkbox"/>
(b) Breech presentation.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Klippel-Feil syndrome.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Vestibular imbalance.	<input type="checkbox"/>	<input type="checkbox"/>
 <b>Question 3:</b> Which of the following statement(s) regarding Monteggia treatment of congenital radioulnar synostosis is/are false?		
(a) Treatment recommendations are made based on cosmetic appearance.	<input type="checkbox"/>	<input type="checkbox"/>
(b) Surgery is needed if the pronation deformity is more than 60°.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Bilateral deformities with less than 30° of pronation generally do not require surgery.	<input type="checkbox"/>	<input type="checkbox"/>
(d) The only surgical option is corrective osteotomy.	<input type="checkbox"/>	<input type="checkbox"/>
 <b>Question 4:</b> Which of the following statement(s) regarding congenital trigger thumb is/are true?		
(a) Trigger thumb is due to thickening and constriction of the fibrous extensor sheath that surrounds the extensor pollicis tendon at the base of the thumb.	<input type="checkbox"/>	<input type="checkbox"/>
(b) Treatment is always surgical.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Radiographs are needed to confirm the diagnosis.	<input type="checkbox"/>	<input type="checkbox"/>
(d) Surgery involves releasing the A1 pulley overlying the flexor tendon.	<input type="checkbox"/>	<input type="checkbox"/>
 <b>Question 5:</b> Which of the following statement(s) regarding cubitus varus deformity is/are false?		
(a) A common cause is malunion of a supracondylar fracture of the humerus in children.	<input type="checkbox"/>	<input type="checkbox"/>
(b) It results from inadequate reduction or failure to maintain reduction.	<input type="checkbox"/>	<input type="checkbox"/>
(c) Corrective supracondylar osteotomy is performed for functional loss.	<input type="checkbox"/>	<input type="checkbox"/>
(d) An untreated cubitus varus deformity may increase the risk of lateral condyle fracture and tardy ulnar nerve palsy.	<input type="checkbox"/>	<input type="checkbox"/>

**Doctor's particulars:**

Name in full: \_\_\_\_\_

MCR number: \_\_\_\_\_ Specialty: \_\_\_\_\_

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**Submission instructions:****A. Using this answer form**

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2. Indicate your responses by marking the "True" or "False" box
3. Fill in your professional particulars.
4. Post the answer form to the SMJ at 2 College Road, Singapore 169850.

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1. Log on at the SMJ website: URL <<http://www.sma.org.sg/cme/smj>> and select the appropriate set of questions.
2. Select your answers and provide your name, email address and MCR number. Click on "Submit answers" to submit.

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1. Answers will be published in the SMJ October 2006 issue.
2. The MCR numbers of successful candidates will be posted online at <http://www.sma.org.sg/cme/smj> by 15 October 2006.
3. All online submissions will receive an automatic email acknowledgment.
4. Passing mark is 60%. No mark will be deducted for incorrect answers.
5. The SMJ editorial office will submit the list of successful candidates to the Singapore Medical Council.