TEST YOUR KNOWLEDGE

No thumbs up for the boy!

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Case summary

A 7-year-old boy presented with both of his thumbs flexed (Figure 1), the inability to perform the 'thumbs-up' gesture, and difficulties in opposing thumbs and fingers in activities such as holding a pencil during writing, pincer movements, and picking up small objects. His mother only observed the deformity for one month after the boy complained of difficulty in writing during class. He had no prior history of trauma to the thumbs or pain and swelling in other joints. There were no other birth anomalies noticed by his parents. Upon examination, the boy had flexion deformities of the interphalangeal joint (IPJ) on both of the thumbs. Passive range of motion of the IPJ was not possible and both were fixed at 60° of flexion. Power of both thumbs could not be assessed as there was no motion of the joint. The range of motion of the metacarpophalengeal joints (MCPJ) of both thumbs and all other finger joints was normal. A 0.5cm x 0.5cm subcutaneous nodule was palpable at the volar crease of the MCPJ bilaterally. The nodule was tender on palpation; however, no overlying skin changes were evident.



Figure 1. Clinical picture showing bilateral thumb IPJ flexion deformity.

Questions

- 1. What is the most likely diagnosis?
- 2. What are the differential diagnoses?
- 3. What is the natural history of the condition?
- 4. What are the treatment options?

Answers:

1. The most likely diagnosis is bilateral congenital trigger thumb. As there is no history of trauma to the thumbs, it is most likely that the deformity has existed since birth but went unnoticed by the parents until one month prior to presentation. As is usually the case, the child only sought treatment after a functional impairment became noticeable, i.e., difficulty in writing. Congenital trigger thumb occurs in 3.3 cases per 1000 live births and is caused by a disturbance in normal flexor pollicis longus (FPL) tendon gliding due to a size mismatch between the tendon and A1 pulley. This mismatch occurs when there is fibrous tissue formation on the FPL tendon of the thumb within the A1 pulley.¹ The condition is characterized clinically by "triggering" or an inability to fully extend the IPJ of the thumb. The definitive etiology is unknown, but studies suggest dysfunctional myofibroblast may be responsible for the condition. Congenital trigger thumb is rare,² and some authors suggest the term congenital is a misnomer as it does not necessarily manifest at birth.³

2. The differential diagnoses of flexion deformity of the thumb in a child include congenital clasp thumb and thumb contracture secondary to trauma. These can be readily excluded by a history and physical examination. Congenital clasp thumb, also known as the 'thumb-inpalm' deformity, is a flexion deformity due to the contracture of the MCPJ, as opposed to a flexion deformity at the IPJ of the thumb. Furthermore, congenital clasped thumb is commonly associated with systemic conditions, such as cerebral palsy and arthrogryposis, whereas congenital trigger thumb occurs typically in isolation, without any other conditions or syndromes. No further investigation is required as the condition can be diagnosed clinically. Radiograph of the hand is useful in cases

with suspected metacarpal or phalanx fractures.

- 3. Spontaneous resolution can be expected in up to 63% of cases but may take up to 4 years in duration.⁴ In a retrospective review of 31 trigger thumbs in 23 children with a mean age of 7.46 years, surgical release of the A1 pulley was successfully achieved in all patients, and there were no recurrences or complications noted at the average follow-up period of 2 years and 3 months. The author also concluded that a satisfactory result can be expected after surgical release of the A1 pulley to facilitate FPL tendon gliding in trigger thumb, even in delayed diagnosis or late treatment.⁵
- 4. Passive range of motion exercises performed by the parents may help to hasten the spontaneous recovery of the trigger thumb. However, in cases which cause a disturbance in functional daily activities, have bilateral involvement, and are seen in children more than 4 years old, early referral for surgical release in advised.

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