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Distal humeral epiphyseal separation

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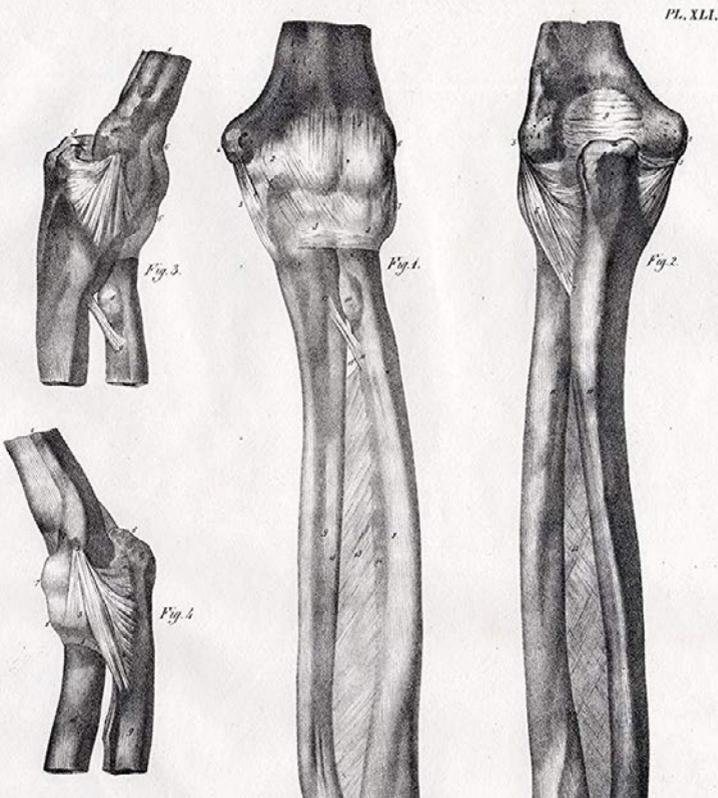
Abstract: Distal humeral epiphyseal separation is a rare orthopedic condition of the newborn (incidence 1:35'000) that requires immediate surgical correction. Untreated, the condition can lead to elbow cubitus varus deformity or dysfunction (1, 2). Diagnosis is challenging as the condition can be easily overlooked or misinterpreted as a brachial plexus palsy or an elbow dislocation. We present the case of a newborn patient whom we treated in multidisciplinary collaboration with our colleagues at the Department of Pediatric Orthopedic and Trauma Surgery and Radiology at the University Children's Hospital of Zurich.

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Distal humeral epiphyseal separation



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INTRODUCTION

Distal humeral epiphyseal separation is a rare orthopedic condition of the newborn (incidence 1:35'000) that requires immediate surgical correction. Untreated, the condition can lead to elbow cubitus varus deformity or dysfunction (1, 2). Diagnosis is challenging as the condition can be easily overlooked or misinterpreted as a brachial plexus palsy or an elbow dislocation.

We present the case of a newborn patient whom we treated in multidisciplinary collaboration with our colleagues at the Department of Pediatric Orthopedic and Trauma Surgery and Radiology at the University Children's Hospital of Zurich.

CASE REPORT

This boy was born at 38 0/7 weeks gestation by elective Cesarean section and adapted well with Apgar scores of 8, 9 and 9 at 1, 5 and 10 minutes, respectively. His birthweight was 3'750 g (P75–90), length 51 cm (P25–50) and head circumference 36 cm (P75–90).

Clinical examination on the first day of life (DOL) revealed moderate swelling of the right upper arm as well as reduced range of motion and obvious pain on movement of the affected limb. Finger movements were intact and the entire arm was warm and well perfused. We initially suspected a traumatic cause for the symptoms, but no delivery complication had been reported, and we did not identify any hematomas. Our clinical examination with respect to posture and movements was not consistent with a brachial plexus palsy either.

Plain X-ray investigation of the right arm on DOL 2 did not point towards a definite diagnosis (Fig. 1). Subsequent ultrasound (US) examination of the right elbow on DOL 4 revealed a dislocated separation between the humeral epiphysis and humeral diaphysis and confirmed the diagnosis of distal humeral epiphyseal separation (Fig. 2). Given this finding, we were able to discern a malalignment of the ulna and radius in relation to the humerus on the previously obtained X-ray image. This observation has been described as pathognomonic and can point towards the diagnosis of distal humeral epiphyseal separation (1, 2).



Fig. 1

Preoperative plain antero-posterior radiograph of the right arm in supination showing medial displacement of the proximal radius and ulna with respect to the distal humeral metaphysis (asterisk).

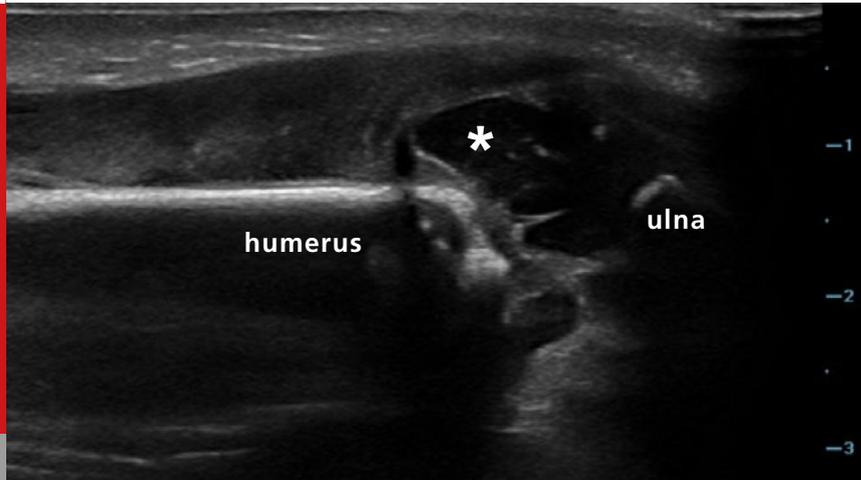


Fig. 2

Preoperative posterior longitudinal ultrasound view of the right elbow showing the distal humeral epiphyseal separation (3) (dislocated humeral epiphysis).*

The treatment consisted of percutaneous pinning in conjunction with intraoperative arthrography (DOL 8). This technique allowed for visualization of the epiphysis, which is invisible on plain radiographs, and confirmation of the correct position after reduction and fixation (Fig. 3). The arm was immobilized with a plaster of Gilchrist, a soft detachable bandage that holds and immobilizes arm and shoulder.

The pins were removed three weeks after surgery (Fig. 4). Postoperative follow-up at two months and one year of age showed early restoration of function and abundant bony healing. Arm movements were unrestricted with symmetrical function and normal alignment of the elbow. The next follow-up is planned at the age of three years.



Fig. 3

Intraoperative arthrogram to confirm anatomic reduction and visualize the epiphysis; the fixation pins and distal humeral epiphysis are visible in the correct position (arrow) (4).

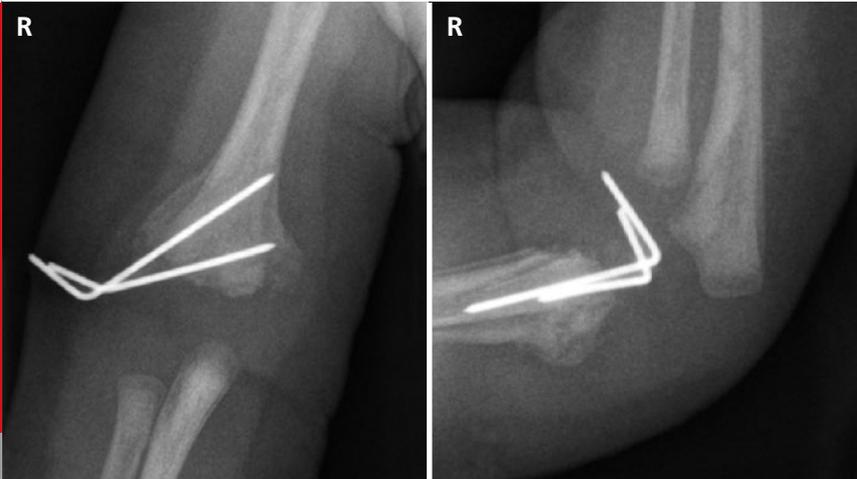


Fig. 4

Follow-up three weeks after surgery with correct consolidation of the distal humerus.

DISCUSSION

Distal humeral epiphyseal separation is a rare condition that mainly occurs in children younger than two years before the distal humeral epiphysis is ossified (5). It seems to be caused by traumatic insults with excessive traction and rotation of the forearm, and can be a consequence of traumatic delivery.

Brachial plexus palsy, elbow dislocation and fracture of the humerus or clavicle are the main differential diagnoses (Table 1). The different clinical appearances are discussed in the subsequent section.

Differential diagnoses

Distal humeral epiphyseal separation

Brachial plexus palsy

- Erb-Duchenne's palsy, C5-C6
- Klumpke's palsy C7, C8-T1

Elbow dislocation

Clavicle fracture

Humerus fracture

Parrot's pseudo-paralysis caused by congenital syphilis

Table 1. Main differential diagnoses of a hypokinetic painful upper extremity.

Brachial plexus palsy is usually associated with a typical history of obstructed labor, complicated by shoulder dystocia with lateral neck flexion and unequivocal clinical signs. Brachial plexus lesions are caused by stretching of the nerve fibers, leading to a neurapraxic injury or even nerve rupture. Usually, the upper nerve fibers are affected (C5 and C6, so-called Erb's palsy), leading to loss of function of shoulder and biceps muscles. The palmar grasp reflex is intact. Affected newborns typically keep the arm internally rotated and pronated in a limp position by the side of the body (also known as waiter's tip position). They are unable to abduct or externally rotate the arm, or supinate the forearm (Fig. 5). If there is phrenic nerve involvement, concurrent diaphragmatic palsy may occur, with elevation of the ipsilateral hemidiaphragm and respiratory distress manifesting as see-saw breathing.



Fig. 5

Left-sided Erb-Duchenne's palsy in a three-day-old term infant.

Klumpke's palsy describes a lower trunk injury involving the C8 and T1 nerve roots with or without involvement of C7. It is rare and usually combined with an upper plexus palsy. It affects the intrinsic muscles of the hand and the flexors of the wrist and fingers. Depending on the type of injury to the brachial plexus, symptoms vary from mild to severe. The affected forearm is flaccid with the hand held in the claw position. The palmar grasp reflex is absent (Fig. 6). When sympathetic nerve fibers (T1) are involved, a Horner's syndrome with miosis, ptosis and enophthalmos is present.



Fig. 6

Left-sided Klumpke's palsy in a two-day-old term infant.

Elbow dislocation is usually a painful traumatic birth injury but can rarely also be due to a congenital malformation with incomplete development of the humerus or ulna. In such cases, the elbow joint is unstable and can easily dislocate during the process of delivery. Concomitant injury to the radial artery can lead to peripheral circulatory disorders. Clinically the elbow is deformed and swollen, and the range of motion is restricted.

An isolated clavicular fracture (Fig. 7) is either due to difficult shoulder delivery or caused by an outstretched arm during birth. Humerus fracture (Fig. 8) can occur after breech delivery and following delivery of the arm. Clavicular fracture is often diagnosed as an incidental clinical finding with palpable callus formation. Restriction of movements by pain result in asymmetrical movements of arms and shoulders. As in the case of humerus fracture, swelling, hematoma, anatomical deformities, possibly crepitation and also pain-related restriction of movements can be observed. The diagnosis of a humerus fracture can be confirmed with a plain radiograph.

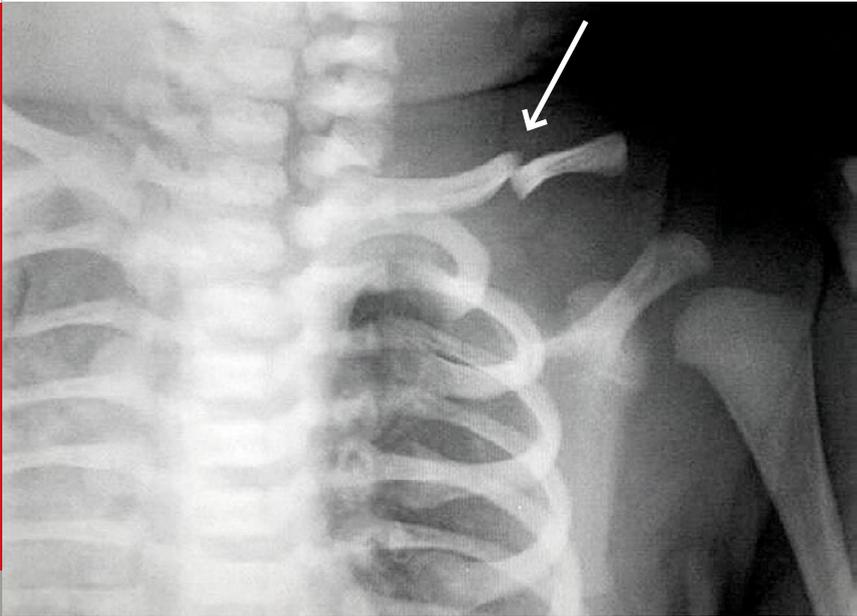


Fig. 7

Left-sided clavicular fracture (arrow).



Fig. 8

Left-sided humerus fracture (arrow).

Parrot's pseudo-paralysis caused by congenital syphilis is not a paralysis in the traditional sense of the word. Rather, it represents an osteochondritis with periostitis of the upper arm, which causes a pain-induced reluctance to move the arm. Two thirds of newborns with congenital syphilis are asymptomatic. Further possible early manifestations are intrauterine growth restriction, anemia, hydrops, hepatosplenomegaly, lymphadenopathy, palmar and/or plantar rash, bullous lesions, saddle nose deformity, and Hutchinson's triad, i.e. interstitial keratitis and chorioretinitis, 8th cranial nerve palsy and later in life characteristic teeth malformations (Hutchinson's incisors and mulberry molars). Further on in the course of the illness, periosteal fissures, mental retardation and seizures can occur, as well as tabes dorsalis after the age of ten years.

In the present case of a newborn with a painful hypokinetic limb, the clinical presentation and radiographic findings were first considered inconclusive. In neonates, the epiphysis is radiologically invisible, which makes epiphyseal lesions difficult to recognize. An important diagnostic tool in the assessment of humeral epiphyseal separation is the radiographic exam of the forearm in supination in order to interpret the alignment of the ulna in relation to the distal humeral metaphysis. Ultrasonography is useful to visualize the epiphysis and to confirm the diagnosis of a distal humeral epiphyseal separation (Fig. 2) (2).

Because of the rare nature of this injury, the optimal treatment is not well established. Incorrect reduction of the displaced epiphysis will frequently lead to cubitus varus deformity or dysfunction. For this reason, there is general agreement on rapid surgical correction of this pathology (1, 6). The flexed position required for reduction is not ideal for visualizing the fracture fragments by ultrasound. Direct elbow arthrogram is a useful alternative that reveals non-ossified structures to confirm anatomic fracture reduction and correct fixation (Fig. 3) (1).

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