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Surgical Management of Coxa Vara in Cleidocranial Dysostosis (Scheuthauer Marie-Sainton Syndrome): Case Report at French Medical Institute for Mothers and Children (FMIC), Kabul, Afghanistan

Shekaib Rahman Behroz[1*], Mirza Mohammad Nijrabi[1] and Ezatullah Faiz[1]

Department of Orthopedic Surgery, French Medical Institute for Mothers and Children (FMIC), Kabul, Afghanistan

[*]**Corresponding Author**: Shekaib Rahman Behroz, Department of Orthopedic Surgery, French Medical Institute for Mothers and Children (FMIC), Kabul, Afghanistan; E-mail: shekaibbehroz@gmail.com

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Abstract

Background: Cleidocranial dysplasia (CCD) is an unusual but well-known autosomal dominant inherited disorder (2,4) and the determinant gene is localized at chromosome 6p21 (8). Mutations in the RUNX2 gene cause this problem, this gen is responsible for osteoblast differentiation and also it is important for both membranous and enchondral ossification (8, 5).

Cleidocranial dysostosis was first described by Marie and Sainton in 1898. It is also known as Marie and Sainton disease. Patients may have excessive movement and motion of the shoulder as clavicles are not developed fully. Late closure of the fontanels, open skull sutures and multiple wormian bones are the other finding. Due to having short ribs the thoracic cavity seen like a small Bell shape (1). It is a disorder or abnormality of bones formed by intramembranous ossification (primarily the clavicles, cranium, and pelvis). This is a rare anomaly with the incidence rate of 1/1,000,000, without sex or ethnic group predilection (3). Differential diagnoses of CCD comprise of Yunis-Varon syndrome, hypophosphatasia (perinatal and infantile forms), osteogenesis imperfect congenita (type 1) and clavicle congenital pseudarthrosis (7). When there is a positive family history of CCD, prenatal ultrasound can help in early diagnosis by showing an abnormal growth of the clavicles (8).

Objective: The aim of this paper is to describe the outcomes of surgical treatment of Coxa Vara for a child with the Cleidocranial dysostosis at FMIC, Kabul, Afghanistan.

Keywords: Cleidocranial dysplasia, coxa vara, valgization osteotomy

Case Report

A 9-year-old boy presented by his parent to the orthopedic clinic of FMIC with chief complain of abnormal gait, both shoulder, hands and foot deformity. On clinical examination patient had laurching gait, both hip abduction and internal rotation was limited and hypermobility of the shoulder girdles. The approximation of humeral head and hypermobility of shoulders was noticed. The patient had short stature abnormal dental findings, typical facial and skeletal anomalies of the skull and clavicle are pathognomic of Cleidocranial dysplasia and the diagnosis of CCD was established.

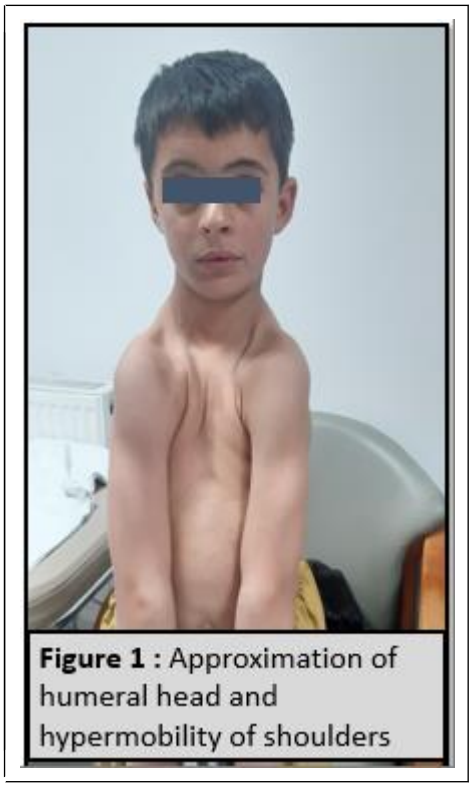

Figure 1 : Approximation of humeral head and hypermobility of shoulders

Investigation

The patient was then referred to radiology department of FMIC. On radiographic evaluation the following findings were noted: Pelvic x-ray shows both side increased Hilgenreiner-epiphyseal angle (more than 60 degrees), decreased femoral neck - shaft and femoral head-shaft angles which confirm the diagnosis (bilateral coxa vara), and a combination of delayed ossifcation of the ischium and pubis.

Chest x-ray reveals both side hypoplastic clavicular and scapula (Figure 3). Skull radiograph shows multiple Wormian bones (Figure 8). Hand x-ray shows accessory epiphysis and short hypoplastic distal phalanges in both sides (Figure 2).

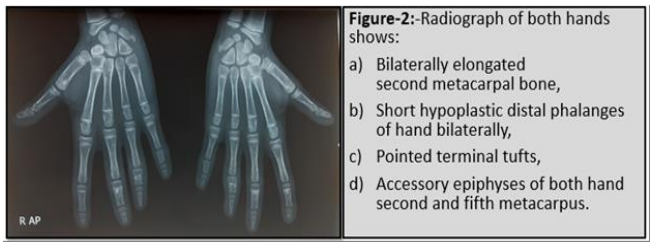

Figure-2:-Radiograph of both hands shows:

a) Bilaterally elongated second metacarpal bone,
b) Short hypoplastic distal phalanges of hand bilaterally,
c) Pointed terminal tufts,
d) Accessory epiphyses of both hand second and fifth metacarpus.

Figure 2: Radiograph of both hands:

a. bilaterally elongated second metacarpal bone

b. Short hypoplastic distal phlanges of hand bilaterally

c. pointed terminal tufts

d. accessory epiphyses of both hand second and fifth metacarpus

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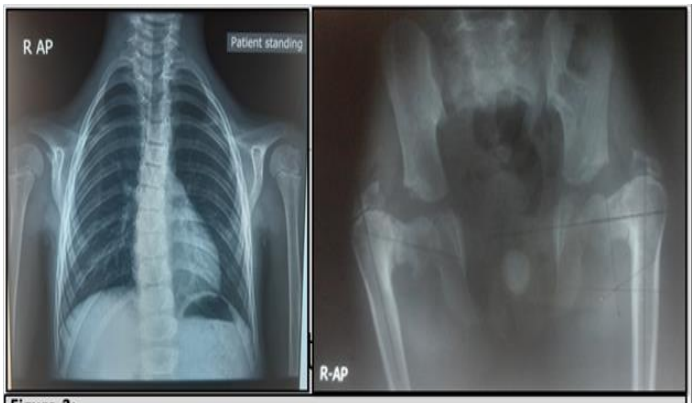

Figure-3:

a) Chest x-ray shows: Both side hypoplastic clavicular and scapula.

b) Pelvic x-ray shows: Increased Hilgenreiner-epiphyseal angle, decreased femoral neck - shaft angle and femoral head-shaft angle which confirm the diagnosis (bilateral coxa vara), and a combination of delayed ossifcation of the ischium and pubis.

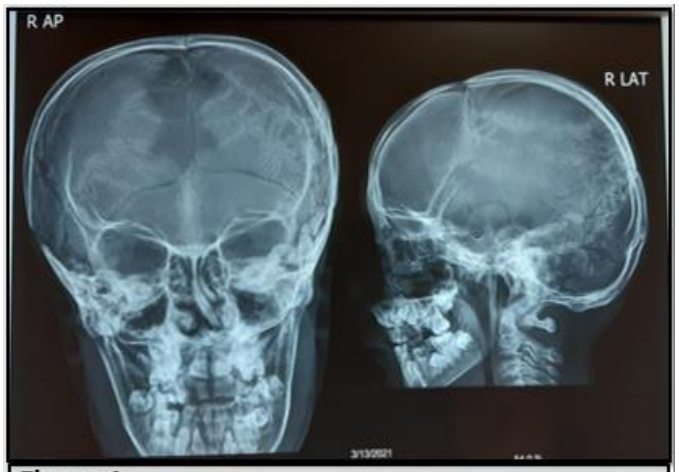

Figure-4:
Skull radiograph showing multiple Wormian bones.

Procedure

Under caudal anesthesia and supine position on the orthopedic table and under the guide of fluoroscopy by one incision through the lateral part of proximal thigh the site exposed Valgization osteotomy (intertrochanteric valgus osteotomy) has been done for right side hip and fixed with five-hole plate and child was immobilized with hip pica cast for two months, after two months spica cast is removed physiotherapy advised and regular follow up done (Figure-5). 8month after first surgery we did same procedure for right side hip. Now 5month post second operation the child has normal hip movement and gait, femoral neck - shaft angle and femoral head-shaft angles are within normal range (Figure-6).

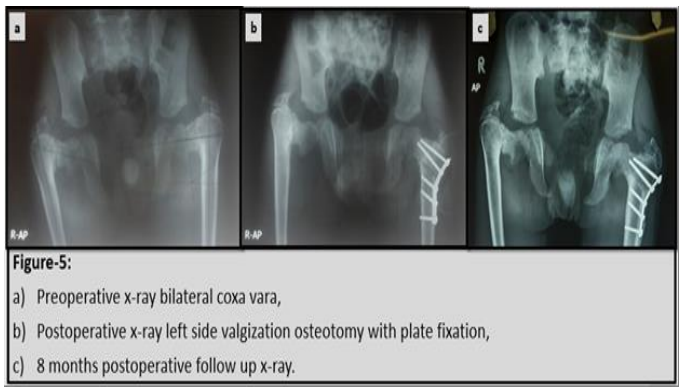

Figure-5:

a) Preoperative x-ray bilateral coxa vara,
b) Postoperative x-ray left side valgization osteotomy with plate fixation,
c) 8 months postoperative follow up x-ray.

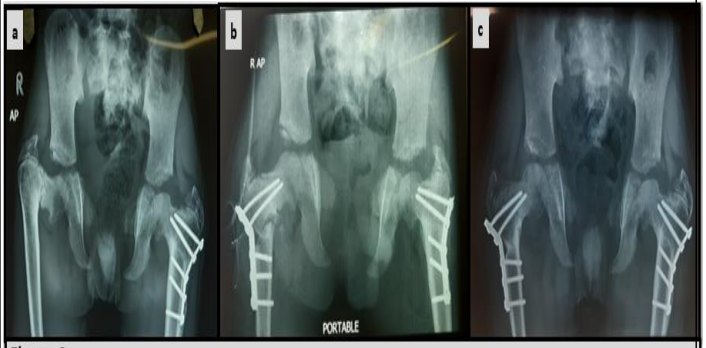

Figure-6:

a) X-ray bilateral coxa vara(operated Left side),
b) Postoperative x-ray both side valgization osteotomy with plate fixation,
c) 5 months postoperative follow up x-ray for Right side.

Coxa vara management for cleidocranial dysostosis

Conservative management for coxa vara in CCD patient is not beneficial and surgical treatment is the gold standard. In general, surgery is required to promote ossification of the femoral neck and correct the deformity. The exact and correct surgical indications for coxa vara include neck-shaft angle less than 90°, Hilgenreiner's epiphyseal angle greater than 60°, and documented progression of worsening of varus angulation. The exact timing of surgery has not been unanimously determined but there is some document which shown recurrence in children

for whom surgery performed before age of 6 years. The optimal age must be determined individually, to prevent early closure of the capital femoral epiphysis and subsequent leg length discrepancy and greater trochanteric advancement the child should not be too young. Similarly, the child must also not be too old, so that enough time remains for development of growth before growth plate closure (8).

Conclusion

In order to make a diagnosis for CCD the finding of the characteristic facial, cranial and clavicular deformities are necessary. It is essential to recognize hip abnormality early, especially bilateral coxa vara, in order to avoid deformity and delay in surgical management. We report the surgical management of coxa vara for a case of Cleidocranial dysplasia because of its rarity.

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