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Management of coxa vara in congenital femora deficiency

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Abstract
Introduction: Congenital femoral deficiency (CFD) is a rare condition, with various classifications. Few have reported on the valgus derotation osteotomy for the correction of coxa vara deformity in CFD. We report our experience in coxa vara correction in cases of congenital femoral deficiency.
Patients and Methods: Six cases with congenital femoral deficiency with the bone abnormality presented in the form of femoral shortening with coxa vara without acetabular dysplasia, cases were classified according to Paley's (only CFD type 1 were included). Cox avara correction was done using a locked proximal femoral plate with femoral shortening and soft tissue release. Average follow-up was 13 months. Evaluation of the outcome was done radiologically and clinically using the Children's hospital Oakland hip evaluation scale (CHOES)
Results: Radiographic union was achieved in all cases at 6 weeks follow-up as evident by bridging callus on at least 3 cortices. CHOES score was excellent in 2 cases and good in 4 cases. We had no significant complication in any of the cases at final follow-up.
Conclusion: Valgus derotation osteotomy with adequate shortening and soft tissue release for the correction of proximal femoral varus in CFD using locked is safe, reliable with minimal complications.

Keywords: Valgus derotation- CFD- Congenital femoral deficiency

Introduction
Congenital femoral deficiency (CFD) is a rare condition with an incidence of one in 50000 to 200000, mainly occurring as a sporadic condition [1-3]. There are many varieties of this condition, with various classifications, from Aitkin in 1959 to Paley in 1998. It was originally classified as a type of coxa vara [2, 4, 5]. Is mostly a proximal deficiency with varus neck shaft angle and variable degrees of ossification [5-10].
Few have reported on the valgus derotation osteotomy for the correction of coxa vara deformity in CFD, which was done using cannulated blade plate [11] or rush rod and tension band construct [4]. Recurrence of the deformity has been observed after correction, which may be caused by asymmetrical growth of the proximal femur [12]. Thus, soft tissue release is required in all cases to correct the NSA without tension, to avoid deformity recurrence [11]. We report our experience in coxa vara correction in cases of congenital femoral deficiency.

Patients and Methods
This is a prospective study of 6 cases with congenital femoral deficiency with the bone abnormality presented in the form of femoral shortening with coxa vara without acetabular dysplasia, cases were classified according to Paley's and operated upon in the period between 2020-2023.
Inclusion criteria were unilateral immature patients (before physeal closure), with CFD type 1 according to Paley's classification. Cases with previous hip surgery were excluded. All cases underwent informed consent, preoperative evaluation is done using radiographs to evaluate the classification and bone abnormality (Fig. 1), and MRI in cases showing delayed ossification in the neck to exclude pseudarthrosis.

Surgical method

Patients were placed in the supine position, with pillow placed under the ipsilateral buttock. Lateral incision to the proximal femur was utilized, with proximal extension to the iliac crest, soft tissue starts with the release of the iliotibial band from the tensor fascial lata, rectus femoris and iliopsoas tendon at the pelvic brim, tight sartorius fascia is also released, and the abductor muscles are released from their attachment to the iliac crest by an apophyseal splitting approach.

Using a locked proximal femoral pediatric plate with 130 degrees angle, the guide wire is placed in the center of the head in the AP and lateral view (Fig. 2 A). The proximal screws are placed in the neck.

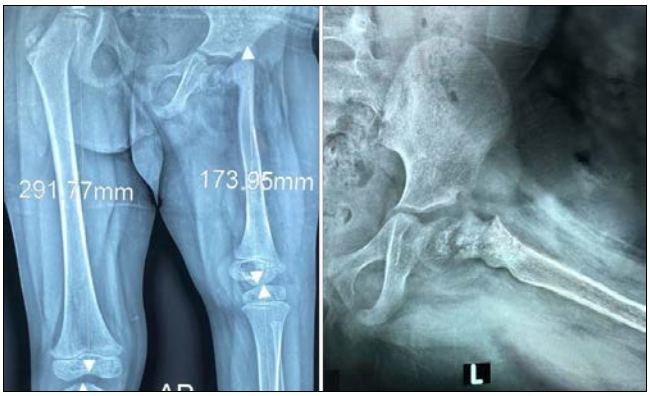

Fig 1: Anteroposterior and lateral view of the femur for preoperative evaluation

The Site of osteotomy was placed at the bend of the plate, which was done transversely using a power saw (Fig. 2 B). To perform valgus-derotation osteotomy properly, It is important to release the quadriceps circumferentially and the linea aspera, allowing the proximal and distal ends to overlap. The amount of overlapped bone is resected, the distal end of the plate is fixed to the femoral shaft, with correct anteversion of about 15 degrees (Fig. 2 C), this is measured using the central guide of the plate (which is central in the neck), and the trans-epicondylar axis in the knee.

Fig 2: (A) insertion of the plate guide wire (B) insertion of the proximal screws (C) plate fixation to the distal segment

Closure includes suturing back the iliac apophysis after cutting the protruding part of the iliac crest to allow tension free closure, tensor fascia lata is sutured to the greater trochanter, subcutaneous tissue and skin are closed over a drain and a one and half spica cast is applied.

Post-operative care

Spica remained for a period of 6 weeks, followed by removal and start weight bearing. Evaluation of the outcome was done radiologically (using Anteroposterior and lateral x-rays (Fig. 3) and clinically using the Children's hospital Oakland hip evaluation scale (CHOES) [12] for evaluation of the range of motion (Table 1).

Fig 3: Anteroposterior and lateral view at 12 months follow-up

Table 1: Hip range of motion was classification based on the Children's Hospital Oakland Hip Evaluation Scale (CHOES)

Hip ROM	Full range	Slight limitation	Moderate limitation	Severe limitation
Internal rotation	>39	39-30	29-16	<16
External rotation	>39	39-30	29-16	<16
Hip flexion	>114	101-114	90-100	<90
Hip Abduction	>39	39-30	20-29	<20

Results

The cases included in this study were classified according to paley's classification into 1A3 (3 cases), 1B1 (2 cases), and one case was 1B3. 4 cases were right sided 2 cases. There was three males and three females with average age 4.8 SD 1.6 years.

Follow-up period averaged 13 months (1.6). Average preoperative neck shaft angle 66.5 degrees (10.7). Average postoperative neck shaft angle 128.8 (7.2).

Radiographic union was achieved in all cases at 6 weeks follow-up as evident by bridging callus on at least 3 cortices. CHOES score was excellent in 2 cases and good in 4 cases. We had no significant complication in any of the cases at final follow-up. There was no significant correlation between age, sex, type of CFD, and the final clinical or radiological outcome.

Discussion

Congenital femoral deficiency (CFD) is relatively rare, with many variable presentations. It mainly presents with coxa vara with or without acetabular dysplasia or pseudarthrosis [2, 7]. There are many classifications for this condition, however, the Paly classification is both expansive and guided the management [9].

The general recommendations is to correct the coxa vara to at least 120 degrees in cases of CFD [13]. Shortening is necessary to correct the coxa vara to a normal neck shaft angle to decrease soft tissue tension. Our average pre-operative neck shaft angle was 66.5 degrees while average postoperative Neck shaft angle was 129 degrees. Eidelman *et al.* [2] corrected the NSA from an average 92 degrees pre-operatively to a post-operative angle that was an average of 115 degrees. Grill *et al.* [14] performed hip stabilizing procedures in patients by performing valgus derotation osteotomy but did not provide pre or post-operative values for NSA.

The clinical outcome based on the CHOES score was

excellent in 2 cases and good in 4 cases, with no significant complication in any of the cases at final follow-up. All cases achieved union at the osteotomy site with no need of bone grafting. Pseudarthrosis of the subtrochanteric region is removed as an apart of the shortened segment, Paley type 2 with frank pseudarthrosis was not included in our study as this condition is more severe and requires more extensive surgical technique and fixation [11].

Soft tissue release is required in all cases to correct the NSA without tension, and to avoid deformity recurrence, which may be caused by asymmetrical growth of the proximal femur [12]. This mostly constituted of flexion and abduction contracture release, [2] however, reports for soft tissue release performed for CFD have been heterogeneous [15].

In our series, abduction contracture release was done through iliac apophysis split and slide as required during the correction coxa vara. Flexion contracture was released by releasing the rectus femoris and iliopsoas tendon (at the pelvic brim), sartorius fascia and sometimes the sartorius is also released, this was in contrast to Paley's method, where only the fascia covering the sartorius is released [11].

Eidelman *et al*. [2] reported release of hip-flexion-abduction contracture. Hip flexion contracture management was achieved through rectus femoris and tensor release. Abduction contracture management was done releasing the abductors tendon and re-attachment to the greater trochanter prior to lengthening surgery, however, this may result in abductor weakness.

Conclusion

Correction of proximal femoral varus in CFD using locked plate combined with adequate femoral shortening, femoral derotation and soft tissue release is safe, reliable with minimal complications.

Conflict of Interest

Not available

Financial Support

Not available

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