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RESEARCH ARTICLE

MANAGEMENT OF A DELAYED PRESENTATION OF CONGENITAL KNEE DISLOCATION IN A 14 MONTH OLD CHILD WITH LARSEN'S SYNDROME AND PIERRE ROBIN SEQUENCE

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Abstract

Larsen's syndrome is a rare genetic disorder characterized by multiple joint dislocations, abnormal facial features, and ligamentous laxity.Dislocation can involve Hips, knees, elbows, and other joints. it was first described by Larsen et al in 1950 [1]. Pierre Robin sequence is another rare congenital birth defect That is characterized by microganthia, glossoptosis and cleft palate which can lead to variety of functional abnormalities including feeding , breathing and hearing [2,3]. Here is a case report of a14-month-old child with Larsen's syndrome who presented late with multiple joints dislocation including (elbow, hips, knees, and ankle), and failed management of knee dislocation.

Introduction:-

Congenital dislocation of the knee is a rare genetic condition associated with many abnormal structural components, including quadriceps tendon contracture, anterior subluxation of Hamstring tendon, small patella, absent cruciate Ligaments and Tight collateral Ligaments.

Those children usually present with dislocation and femoral condyles can be palpable posteriorly. The spectrum of knee dislocation varies as there are different classification system.

One of which is Tarek classification system that was based on the observation of passive knee flexion compared to the degree of hyper extension and the relation between tibia and femur on x ray [4], figure (1).

In our case the child presented with (Grade 3) ,<30° flexion Clinically and frank dislocation clinically figure (2) and radiographically figure (3).

The child has also presented with manifestation of Pierre robin sequence including, microganthia, and U-shaped cleft palate.

Herewe will focus on the congenital knee dislocation as one of the manifestations of Larsen's syndrome.

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Case Report

A 14 month old boy who had noncontributory family history to Larsen syndrome or pierreRobin sequence, and underwent uneventful intrauterine Life apart from the UTI that the mother had at the 14th week of gestation, for which She was treated with antibiotics for one week.

The baby was born full term at 40 weeks via cesarian section after failed trial of spontaneous vaginal delivery (SVD), as the baby vitals started to disturb.

After birth he was admitted to Neonatal Intensive Care Unit (NICU) for 45 days due to birth asphyxia, with birth weight of 2.5kg.

There he was oncontinuous oxygen therapy without mechanical ventilation for about 30 days, and for 15 days on and off. He was on Nasogastric Tube as well (NGT), as he was diagnosed with Pierre Robin sequence. He was also found to have bilateral abnormal knees and diagnosed with bilateral congenital knee dislocation.

The bilateral knee dislocation treatment was started at day 10 after delivery,using casting method weekly for 8 weeks with no improvement.The mother went to different Hospitals seeking medical Advice.

At presentation to our Hospital the child was found to have abnormal facial features, U-shaped cleft palate, and multiple joint Dislocations including (bilateral Hip, bilateral knee,bilateral ankle,rightradial head subluxation and left radial head dislocation.

On Examination

1. Craniofacial : Abnormal facial features including the small retracted jaw (microganathia) and U shaped cleft palate.
2. Hips: Limited abduction
3. knees: hyperextended with anterior skin folds and palpable femoral condyles posteriorly, and limited knee flexion (Figure 2,3).
4. Ankle: Bilateral dislocation.
5. Elbows: (**Lt** Radial Head subluxation and **Rt**Radial Head dislocation).

At preasentation both hips and knees whre not reducible by closed means.

Tarek CDK grading system

Grade	Range of passive flexion	Radiology
GI	>90°	Simple recurvatum
GII	30–90°	Subluxation/dislocation
GIII	<30°	Dislocation

Figure 1: Tarek grading system of congenital knee dislocation, based on passive range of motion and radiographic findings.

The child was planned for open reduction of bilateral knee dislocationand quadriceps lengthening. hip reduction to be decided later according to the child motor function after physiotherapy.

Operative procedure:

Under general anesthesia the child was put on supine position (figure 4), tourniquet applied bilaterally.

prep and drape was done with aseptictechnique, anterior approach was used, dissection done in layers. The quadriceps lengthening done using V-Y quadricepsplasty.

The reduction was done easily by pressing the distal femoral condyle anteriorly. The Child was found to have no anterior cruciate ligament(ACL) orposterior cruciateligament (PCL) intra operatively. and this could necessitate the need of using knee brace later.

115° and 98° of knee flexion was achieved in the left and right sides respectively, the reduction was maintained with K-wire and the limbs were splinted in flexion (figure 5).

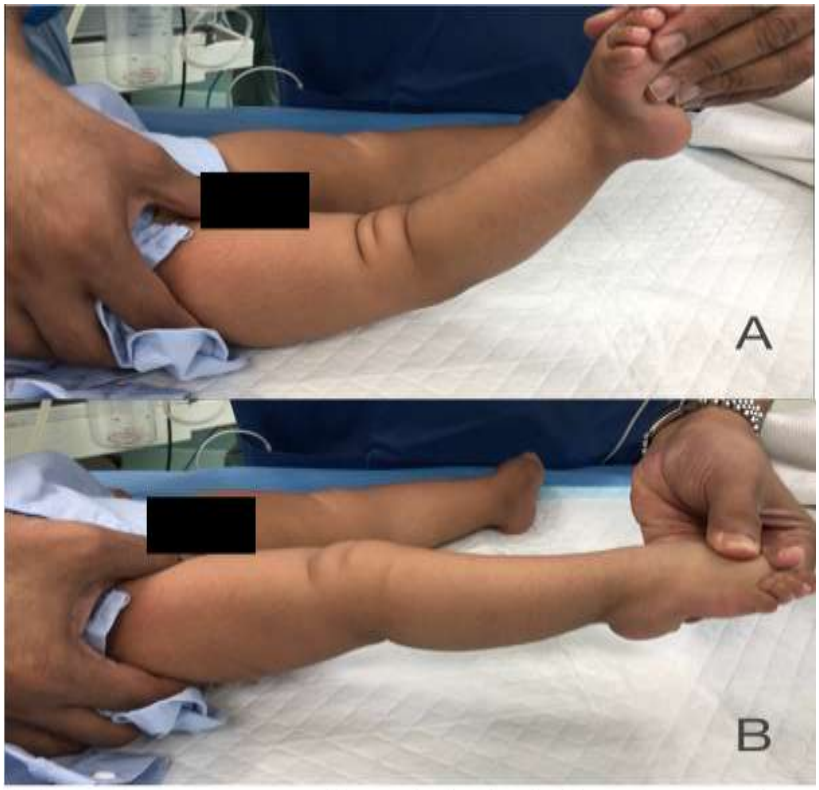

Figure 2: pre operative clinical photos for passive range of motion of the knee (A) extension (B) flexion.

The plan was to change the cast weekly until 90° of flexion at least is achieved bilaterally.

3 weeks post operatively

The k-wires and the splints were removed, and more flexion was gained in both knees (Figure 6). Gradual flexion using weakly cast change was started.

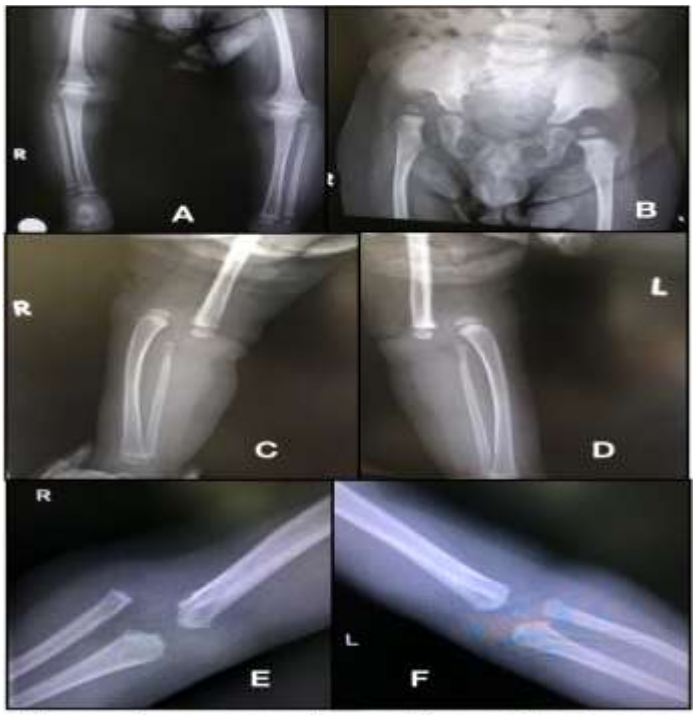

Figure 3: preoperative radiographic Images (A,C,D) knee, (B) pelvis, (E,F) elbows.

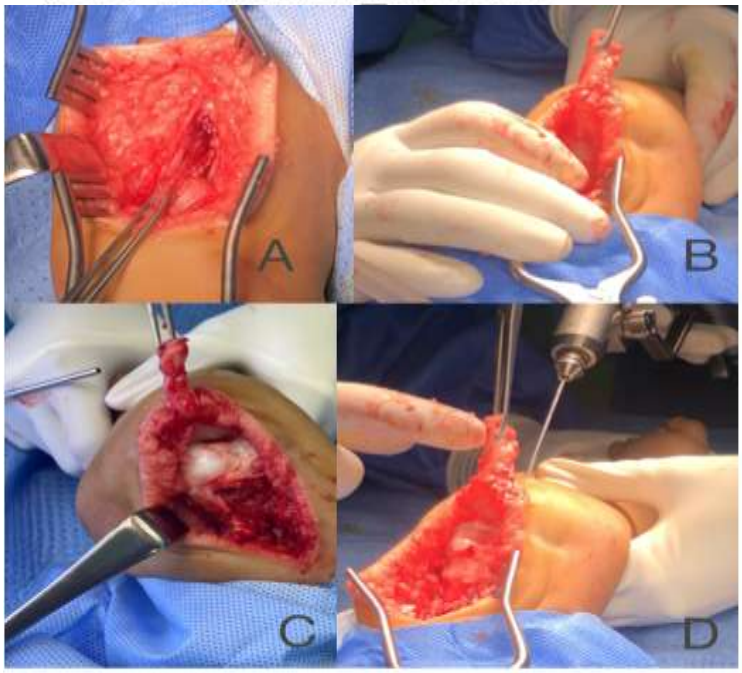

Figure 4: Intraoperative clinical photos of the knee (A) V-shaped, quadricepsplasty, (B) distal part of the quadriceps tendon, (C) post reduction photograph, (D) K-wire application.

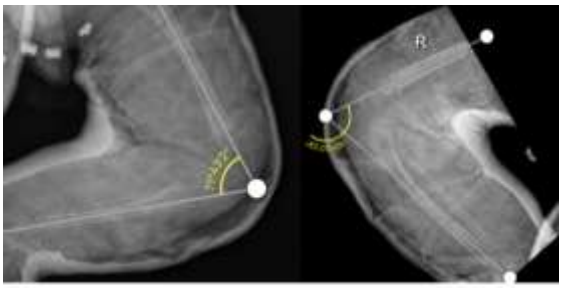

Figure 8 : 6 weeks post operatively more flexion was gained in both knees left knee 71° , right knee 67°.

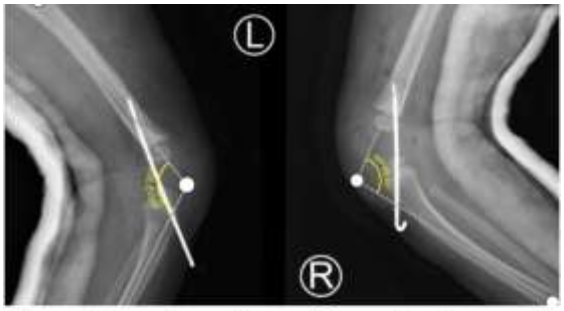

Figure 5: postoperative radiographic Images of the left and right knees after achieving 115°and 98° of knee flexion, in the left and right side respectively, the reduction was maintained with K-wire and the limbs were splinted in flexion.

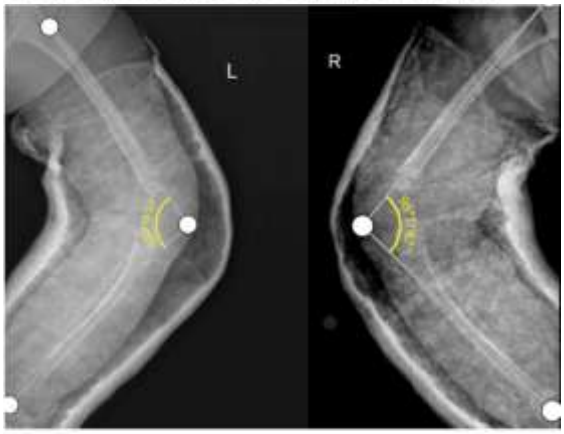

Figure 6 : 3 weeks post operatively , more flexion was gained in both knees left knee 100°, right knee 95°.

6 weeks post operatively

More flexion was gained in both knees, left knee 71°, right knee 67° (Figure 8) . no more flexion was gained during the weekly follow up. hinged knee brace and physiotherapy are then started 2 months post operative (Figure 9).

Figure 7 : 4 weeks post operatively (A) clinical, (B) radiographic.
more flexion was gained in bilaterally left knee 84°, right knee 88°.

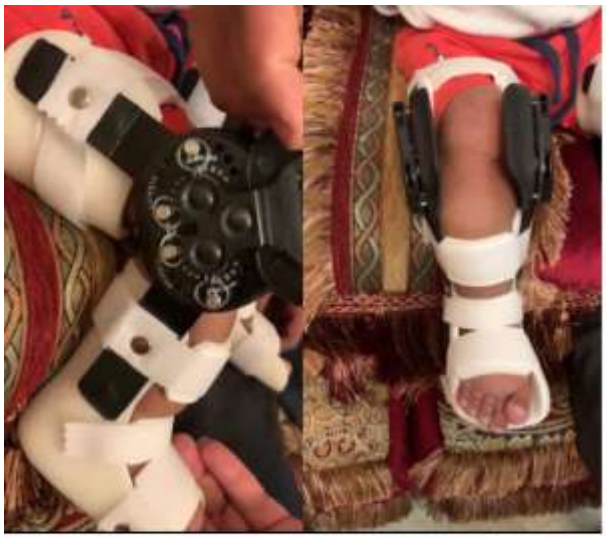

Figure 9 : 2 months post operatively The patient was started on a hinged knee brace bilaterally

2 months later
The child started to stand with support.

Discussion:-

Multiple joint dislocation on the same limb can be challenging in dictating the proper management of each joint dislocation.

Our child has presented with. Bilateral knee, Hip, and Ankle dislocation. We started with the knee first aiming to gain 90 of flexion, via quadriceps Lengthening, open reduction and immobalization on a cast that was changed in weekly mannerfor 9 weeks.In this case there was no rule for nonoperative management alone.

It was mentioned previously that the nonsyndromic knee dislocation usually responds well to non operative management, and has better prognosis than those with multiple dislocations e.g.,Larsansyndrome which represents a challenging situation for planing the management, and can put the tibial metaphysis and epiphysis at risk of plastic deformation [5].

In addition to that, our case has presented with grade 3 knee dislocation,which is an indication for operative management based on Tarek classification [4].

Thereasons why we started with the Knee: 1st is Helping the child to start standing and waking on a brace later, and 2nd is if the child was able to walk he will have the necessary knee range of motion for walking, and This will dictate the necessity for other operative management of the hips and ankles. and3rd is for easier application of spica after open reduction of hip dislocation.

HR Matar and NK Garg has reported a case at an early presentation in patient with bilateral hip dislocation, bilateral knee dislocation and bilateral talipes equanovarus, and they started with Ponseti method for the bilateral talipes and they included the knee attempting closed reduction, and they mentioned that in case of knee and hip dislocation, knee should be addressed first;for easier applications ofhip spica in case of treatment of hip dislocation, However the knee dislocation in their case was treated operatively as the non-operative management failed [6].

Finally, in our case the child started to stand with support, and that dictates the need of planning of open reduction of hip dislocation and correction of ankle deformity.

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