Bilateral Hip Dysplasia in Krabbe Disease

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INTRODUCTION:

Krabbe disease, also known as Globoid cell leukodystrophy resulting from a deficiency in the enzyme galactosylceramidase (GALC) causing extensive demyelination in the central and peripheral nervous systems[1]. In 50% of individuals with Krabbe disease, bilateral hip subluxation has been identified[1].

REPORT:

We report a 6 years old boy with underlying Krabbe disease and epilepsy presented with bilateral hip dislocation. Functionally, patient walked with walking frame. Physical examination noted windswept deformity of the hips with tight hip adductors. Plain radiograph showed right hip subluxation, left hip dislocation and bilateral acetabular dysplasia. Bilateral acetabular index is abnormal with right hip 36 degree and left hip 43.6 degree. Patient underwent bilateral hip adductor release and varus derotation osteotomy (VDRO) with left pelvic Dega acetabuloplasty. Post-operation, both hips were stable and Petrie cast was applied.

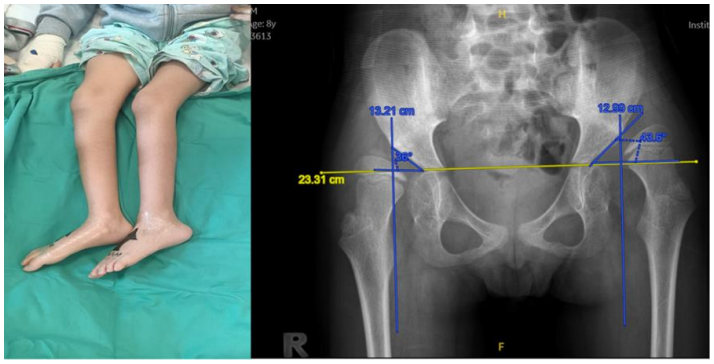

Figure 1: Figure 1: Windswept deformity of the hip (Left) and Pelvis X-ray showing Acetabular index left side 43.6 degree and Right side 36 degree (Right).

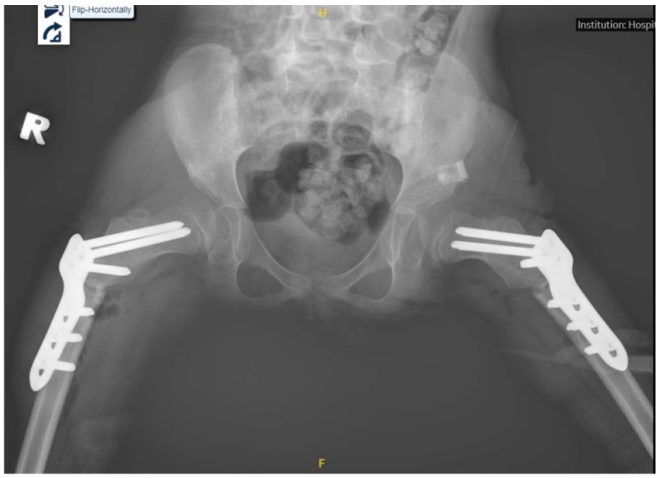

Figure 2: Figure 2: Post operative X-ray post bilateral VDRO and left pelvic Dega acetabuloplasty.

CONCLUSION:

The primary goal in managing dysplastic hip is to achieve and sustain a concentric reduction of the femoral head within the acetabulum, facilitating normal hip development. The Dega acetabuloplasty incorporates benefits from both Salter osteotomy, which involves reorienting the acetabulum, and Pemberton osteotomy, which focuses on reshaping the acetabulum[2]. Femoral osteotomy, involving shortening, derotation, or both, is typically necessary when excessive force is required for hip joint reduction[2]. One-stage procedure entailing open reduction, Dega pelvic osteotomy, and femoral osteotomy are optimum in managing hip dysplasia.

REFERENCES:

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2. Zein A, Khalifa AA, Elsherif ME, Elbarbary H, Badaway MY. Are the outcomes of single-stage open reduction and Dega osteotomy the same when treating DDH in patients younger than 8 years old? A prospective cohort study. *Journal of Orthopaedics and Traumatology*. 2023;24(1):43.