

Case Report of Bilateral Lower Limb Deformity in Paediatric Age Group Secondary to Vitamin-D Resistant Rickets

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Abstract: A 4 years old male child brought to our outpatient department with chief complaint of bilateral lower limb deformity since the child started walking. After thorough clinical examination and investigations, the case was diagnosed as bilateral lower limb deformity secondary to vitamin-D resistant rickets. We managed the case surgically with bilateral epiphysiodesis initially. Later a period of 18 months to 2 years, osteotomy and fixation was done at multiple sites to correct the deformity further. Post operatively both lower limbs were immobilized in above knee pop cast in corrected position for 3 months. Regular follow up was advised. Vitamin-D and calcium supplementation was given throughout the treatment. At regular follow-up, patient obtained a physiological alignment and good bone formation is seen. Corrective osteotomy at different levels and plate fixation and immobilization in corrected position is a good option for the management of lower limb deformities secondary to vitamin-D resistant rickets in children.

Keywords: vitamin-D resistant rickets. Corrective osteotomy. Deformity. Plate fixation

1. Introduction

In 1937 Albright et al first described about vitamin-D resistant hypophosphatemic rickets.¹ This pathology occur in the convoluted tubules of the kidney.² according to some authors this may be due to failure of 1- α -hydroxylation of vitamin D in kidney.³ clinical features include short stature, stunted growth, coxa vara, genu varum or valgum. Early treatment of high dose vitamin D in childhood may prevent bony deformities.⁴ Despite of medical management or delayed presentation; many patients are suffering with deformities and needs surgical correction. We have had experience managing severe deformity in a patient with vitamin-D resistant hypophosphatemic rickets by multiple corrective osteotomies and fixation and immobilization in corrected position.

2. Case report

A 4 Years old male child brought to our outpatient department with chief complaint of bilateral lower limb deformity since the child started walking. Patient was born out of consanguineous marriage. Similar family history was noticed (pt's younger brother). Antenatal, post-natal & birth history was normal. Delayed history of motor milestones present up to 3yrs of age. Immunization as per schedule. Diet history was normal. This child born out of 2nd degree consanguineous marriage.

On examination- Frontal & parietal bossing present Alopecia, Loss of eye brows is there Atrophic shiny skin over entire body like thin plastic membrane. Pectus excavatum, B/L Costochondral beading present. Compensatory scoliosis- present. Widened wrist & Knee joints. Tilted Pelvis. Windswept deformity.

Clinical Images



Investigations- CBP, ESR- Normal, Serum electrolytes- Normal.

LFT- **Increased Alkaline phosphate levels(1600IU/dl)**

serum calcium levels---8.2mg/dl.

serum PTH—68pg/ml.

serum phosphorous levels--3.6mg/dl.

Renal function tests---- Normal.

serum 25(OH) Vit-D—70.4ng/ml.

serum 1,25(OH) Vit-D—61.65pg/ml

CT-Brain- Normal.

X-Rays*Widened physis**Widened epiphysis**Fraying and cupping of metaphysis.**Pencil thin cortex of diaphysis.**Osteopenia*

Opinion was taken from pediatrician and the case was diagnosed as vitamin-D resistant rickets and has been treated with oral calcium supplementation & Vit -D with regular monitoring. After 2yrs **Bilateral Hemi-epiphyseodesis** was done followed by implant removal after 1year.

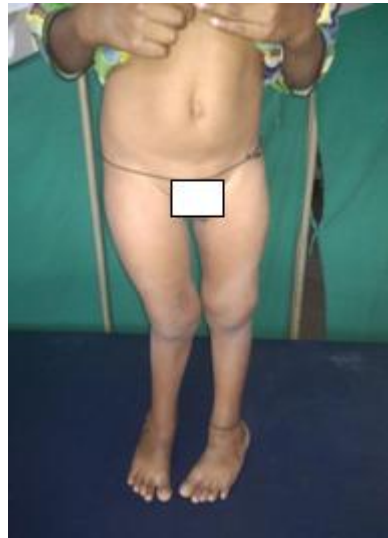
- As the outcome was not satisfactory, we took arthroscanogram and planned for corrective osteotomy at multiple levels.
- We did *Anterolateral close wedge osteotomy of distal femur, at the apex of the deformity on left side and medial open wedge osteotomy at the proximal part of right tibia.*
- Post operatively POP was applied for 3months in corrected position.

Immediate post op3 months follow upIntra-operative Images

Immediate post op x-ray



3 months follow up x-ray



Patient was able to manage his daily routine activities with minimal difficulty.

3. Conclusion

From our result it was concluded that surgical management for deformity of the lower limbs due to vitamin-D resistant rickets by hemi-epiphysiodesis and multiple corrective osteotomy & plate fixation was an effective method with acceptable outcome.

4. Discussion

The syndrome of rickets resistant to normal therapeutic doses of vit-D, but amenable to massive dose was first described by Albright, Butler and Bloomberg in 1927. Very rare **Autosomal recessive** form of rickets with *end organ resistance* to Calcitriol. Caused by mutations in the gene encoding the Vit-D receptor. A unique feature is Alopecia, which is seen in approximately two-thirds of the cases and is the marker of disease severity. Medical management consists of calcitriol and calcium supplementation. Surgical management like epiphysiodesis^{5, 6}, multiple osteotomy and Ilizarov technique⁷ is used.

References

- [1] Albright F, Butler AM, Bloomberg E (1937) rickets resistant to vitamin-D therapy. Am J Dis Child 54:529-547
- [2] Arnstein AR, Frame B (1966) Primary hypophosphatemic rickets & osteomalacia; a review. Clin orthop relate res 49:109-118.
- [3] Rasmussen H, Anast C, Parks J, et al., (1976) 1- α (OH)D3 in treatment of hypophosphatemic rickets. Clin Res 23:486a
- [4] Glorieux FH, Marie PJ, Pettifor JM (1980) bone response to phosphate salts, ergocalciferol, and calcitriol in vitamin-D resistant hypophosphatemic rickets. N Engl Med 303:1023-1031
- [5] Evans GA, Arulanantham K, Gage JR (1980) primary hypophosphatemic rickets. Effect of oral phosphate and vitamin D on growth and surgical treatment. JBJS Am 62:1130-1138
- [6] Mc Alister WH, Kim GS, Whyte MP (1987) Tibial bowing exacerbated by partial premature epiphyseal closure in sex-linked hypophosphatemic rickets, Radiology 162:461-463
- [7] Choi IH, Kim JK, Chung CY et al., (2002) deformity correction of knee and leg lengthening by Ilizarov method in hypophosphatemic rickets; outcomes and significance of serum phosphate levels. J Pediatr Orthop 22:626-631

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