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Ambulatory Rehabilitation of a case of Bilateral Tibial Hemimelia and Right Proximal Focal Femoral Deficiency-

A Case Report

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Abstract

Congenital Limb deficiency with Bilateral Tibial Hemimelia results in deformity and disability of the person not only to the physical body but also psychologically and most of the time we are able to see adult patients with congenital limb deficiency confined to tricycle or dragging themselves on road. Most of them remain underachievers in their life due to their physical and mental disability. We report a rare case of Bilateral Congenital Limb deficiency with Proximal Focal Femoral Deficiency on right side that has been successfully, completed bipedal ambulatory rehabilitation.

Keywords: Tibial Hemimelia, Congenital Limb Deficiency, Proximal Focal Femoral Deficiency

Introduction

Tibial Hemimelia is one of the rarest congenital malformations with a wide range of clinical presentation and it bilateral occurance is most uncommon of the lower limb anomalies. Its estimated incidence is about one in one million live births⁽¹⁾. In majority of cases total absence of tibia occurs and in cases of partial absence distal end is most commonly absent. Tibial Hemimelia is associated with several syndromes such as Werner's syndrome⁽²⁾, LangereGiedion syndrome, tibial hemimelia and split

hand and foot syndrome⁽³⁾ and tibial hemimelia micromelia trigonal brachycephaly syndrome⁽⁴⁾.

Case Report

A 5-year-old female child presented to out outpatient department with deformity of both lower limbs and hands.



She was the first-born child of a Nonconsanginuous marriage. Only positive history in antenatal period was Gestational Diabetes and Hyperemesis. The child was delivered by LSCS.

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Radiograph of limbs taken revealed that she had Bilateral Tibial Hemimelia with Proximal Focal femoral Deficiency and Congenital Dislocation of the Hip on the Right side and Upper limb X ray showed that she had Bilateral Lobster Claw hand. There was delay in motor developmental milestones and was able to sit with support and crawl by 1 year. There is no defect in speech & hearing or vision. She moves around the house by dragging herself and for community ambulation her mother carries her and she was partially dependent on her ADLs (Activities of Daily Living) with a FIM (Functional Independence Measure) Score of 96/126. Hence she was admitted with the Goal of improving her household ambulation and Improvement of the FIM score. Initially the child was trained to balance herself on her knees and walk short distance on her knees following which she was surgically treated with Right Knee arthrodesis and Bilateral adductor tenotomy in collaboration with Orthopedic department. After wound had healed she was given gait training with an Ischial weight bearing Extension type Prosthesis in the Right side and a Bend knee type ischial weight bearing Prosthesis on the Left side.



She was able to ambulate independently using a wheeled walker and her ADLs have improved considerably with a FIM score of 108/126.



Discussion

Classification of Tibial Hemimelia is done using Jone's Classification

- 1. Type-1 a) Total absence of tibia b) Congenital aplasia of tibia with intact fibula.
- 2. Type-2 Proximal tibia is present
- 3. Type-3 Distal tibia is present (Rare)
- 4. Type-4 A divergence of distal tibia and fibula with proximal displacement of talus

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Webber also introduced a score system for Tibial Hemimelia, ranging from zero to 39; the higher the score, the less impairment suffered by the patient. Paley has introduced a new classification system⁽⁵⁾ developed to clarify treatment options and related prognosis. There are 5 types and 11 subtypes with modifiers to better represent Tibial Hemimelia associated deficiencies or duplications.

Reconstructive surgery and prosthesis adapted to growth together with regular post operative follow up are necessary for optimal functional results. If the entire tibia is absent there is subluxation of knee and ankle joints with proximal and lateral positioning of fibula. Severe flexion deformity is also noted in such cases. In such cases Knee disarticulation is generally preferred, although centralization of fibula-Brown's procedure⁽⁶⁾ combined with Syme's amputation has also been described although the long term results are not promising. Nevertheless, there is a lack of evidence about the correct approaches according to the grade of tibial hemimelia. However, development of new surgical options such as hexapod frame offers different options as alternative to amputation $^{(7)}$.

Conclusion

Tibial Hemimelia can be rehabilitated for bipedal ambulation if it is started at an earlier age. This child may require frequent change of prosthesis as her growth progresses and once she has completed growth she may require amputation of the Left foot if she so desires as to correct the cosmetic problem and to give a better prosthesis. By completing the rehabilitation program she has not only improved in her physical abilities but given a renewed confidence to fight against her disabilities.

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