

Ambulating the 'Different': A Prosthetic Approach towards Conservative Management of Congenital Anomalies

Lukeshkumar R. Bhuyar¹, Garima Sharma²

Demonstrator P&O¹, I-MPO²

Department of Prosthetics and Orthotics, All India Institute of Physical Medicine and Rehabilitation, Mumbai, India

Abstract:

Conservative management of persons with congenital anomalies with prosthesis is always a challenging task. In this study we took 4 cases with congenital anomalies and fabricated them a prosthetic fitting. Amputee Mobility Predictor scale has been used to demonstrate functional abilities pre and post prosthetic use by the individuals that contributed to the results. The study outcomes suggest early prosthetic fitting in children resulted into easier adaptation towards corrective rearrangement of body dynamics and improved independency. Adaptive gait in adults demanded profound observation, fine selection of design and components and prolonged gait training. Prosthetic management goals differed in every case but a successful predetermined was achieved.

Keywords: Congenital Anomalies, Prosthetic Rehabilitation, Amputee Mobility Predictor scale, Conservative management, Activities of Daily Living.

Introduction:

Congenital anomalies project a high contribution in infant and early childhood deaths worldwide. According to the world Health Organization, 2,95,000 newborns die in the first month of their lives. In low and middle income countries this condition expresses a painful sight of individual's dependability on family and society throughout lifetime from basic to moderate activities of daily living. In involvement of lower limb, congenital anomalies have altered biomechanical structure and physiology that impairs their gross mobility. Etiology remains idiopathic however there are genetic factors, ionizing radiation, traumas, environmental toxins, critical intrauterine conditions and poor maternal health. In many cases specially remote rural areas, due to lack of awareness or equipped medical assistance the identified type of deformity of anomaly remains uncertain until birth, treatment measure therefore depends upon post natal general health conditions, age and long term functional goals. Advancement in surgery had been solving and supervising management of such conditions and resulted great achievements. Some cases whereas focused on prosthetic arbitration providing a close to normal mobility independence as a disarray limb replacement. Some most common anomalies that often require prosthetic attention are congenital deficiencies of femur, congenital longitudinal deformity of tibia and femur, lower limb transverse deformities and other LLDs. Because there is a wide range of asymmetrical structural appearance, prosthetic management for such cases do not project a consistent prescription and therefore result.

Methods:

Patients reporting to the Out Patient Department of a tertiary rehabilitation and healthcare center were included in this study. Random method of selection was done during the period of September, 2021 to December, 2021 and case studies of 4 patients were documented. A thorough patient history including parent's health history was taken to understand nature of the conditions. Radio graphical images were taken under consideration to evaluate and classify the anomaly. The International Organization for Standardization designation of Transverse and Longitudinal deficiencies of lower limb is been used to conclude diagnosis. After a profound analysis and judgment, treatment goals were organized to provide most effective prosthetic outcomes. Amputee Mobility Predictor scale has been used to demonstrate functional abilities pre and post prosthetic use by the individuals. Gait training and analysis was added as essential tool to study and deduce the presenting case series.

Results:

Case 1

A pediatric case of age 3 years presented with proximal focal femoral deficiency and fibular Hemimelia. Periodic x-ray images confirmed gradual increase in knee contracture mating it an atypical clinical feature. The AMP no PRO score rated 10. First prosthetic fitting was prescribed using LTTP (orfit) casting where metal uprights were added and rotated externally to accommodate LLD and mimic Hip-knee rotation and abduction. An added hind foot rocker was added to the distal unit to decrease initial contact impact force and promote weight transfer. As the young ambulator displayed unapproved biomechanical gait features, another prosthesis was designed. Casting was done in bent knee position and pylon was attached distal to the knee ensuring posterior shift of weight along with COG. The followed AMPPRO score increased to 17 after post fitting gait training.



Case 2

An adult individual of age 26 diagnosed with Proximal Focal femoral deformity of Limb length discrepancy of 7" with functional anatomical knee and ankle joint. Hip extensors and knee flexors were weak, increased lordosis and above average body weight was documented. The AMPnoPRO score was 16. Prescribed prosthesis featured anterior and posterior shell with distal extension and prosthetic foot piece and external rotation of 7 degrees for base of support diagnosed attachment. The weight bearing knee was in 0° flexion and ankle joint 90° alignment. Post gait training the AMP score increases to 21. The gait pattern however demonstrated vaulting of sound limb and hip hiking of the affected side while initiating gait. Energy expenditure from user's perspective had no major differences pre and post prosthetic fitting (subject not willing to give is consent for photographs).

Case 3

A female of age 5 years and 6 months was diagnosed with a rare multiple congenital anomaly at birth. The radiological images concluded distal absence of tibia fibula and complete bony absence of ankle and foot but presence of loose hanging soft tissue. The young individual could ambulate without a prosthesis but it caused extensive flexion of hip knee and foot

dorsiflexion of contralateral limb whereas affected limb ad weak hip abductors and knee flexors in loading. This lowered ambulation also represented inclined transverse alignment of trunk and hip. The AMPnoPRO score was 16, as the subject has adaptive to use of asymmetrical limb. The prosthesis prescribed was patella tendon bearing trans tibial type socket with shank extension and prosthetic foot. The fit was completed in alignment with sound limb and a shorter height of 2 cms in prosthesis for better toe clearance, smooth swing initiation and easier gait. AMP score was documented 26 after the prosthetic fit. Post prosthetic training period was 2 weeks where standing balance was gained in 3 days, aid walking in 8 days and independent walking in 16 days. Patient could easily donn and doff the prosthesis and performed most basic ambulatory tasks quiet efficiently.



Case 4

Another repeating case of a 17 year old male patient, first reported in 2006 after a reconstructive surgery for metatarsal malalignment and syndactly of 3rd 4th and 5th toes resulting into SYMES like amputation. The surgery was performed at the age of 3 after which prosthetic fitting was prescribed. The patient would prefer bare walking and without the prosthesis until age 5, however the limb length did not increase and distal overgrowth of tibia caused disruption and posterior sift of heel pad unfit for distal weight bearing. Patient has been reporting to the hospital ever since and have had 6 prosthesis prescribed until December 2021. The AMPnoPRO score was 12 because of painful weight bearing of affected limb. The recent prosthesis was designed as patella tendon bearing prosthesis with multiaxial foot piece. The later AMP score was 32.



Case 5

A female case of age 20 presented with PFFD in the right limb. Hip ROM was 90 deg. Of flexion, 40 deg. abduction, 20 deg. adduction with full internal and external rotation. The knee presented contracture 30 deg. and full flexion at 140 deg. Ankle had full ROM however the ambulator preferred an equinus weight bearing position. The prescribed prosthesis was designed as ischeal weight bearing socket proximally and metatarsal bearing distally with anterior and posterior shell. AMPnoPRO score was 20 and ambulator could walk both with and without the prosthesis. The ultimate prosthetic goal

here was hand free gait, partial weight bearing at foot and control progressive knee contracture with the prosthesis the AMP score was 26, and unsupported gait could be achieved.



CONCLUSION

Surgical interventions for congenital deformity management has numerous advantageous results assuring close to anatomical functional and structural stability among such individuals. Parents and society of the "different" play a major role in determining future management procedures in particular of those from rural background. The false fear of failed surgery or malpractice and even loss of the 'leftover' limb are some common reasons for not opting surgical procedures.

Prosthetics then provides management with possible assurance of better ADL, improved functional goals and greater self-esteem among individuals. Because every case is a unique presentation by itself despite of a range of classification of diseases available, it is hard to demonstrate standard prosthetic goals. A close study of acquired anatomy and altered biomechanics of their system has to be evaluated to conclude any prosthetic procedure. From selection of materials to providing joints and foot pieces every component can add a significant role of functional ambulation. With the above case studies an improved AMO score was observed post the prosthetic alignment and training. Use of prosthesis to the fullest was a subjective matter for all users as their condition offered them altered choice of walking as in with or without prosthesis. One advantage that could be well understood here is that prosthetic management among people with Congenital Anomalies and Limb Length Discrepancies can benefit by delaying or eliminating secondary physiologic and orthopedic deformities that would otherwise would happen as a result of diverted biomechanics of joints and bone structural alignment.

References:

Bergère A, Amzallag-Bellenger E, Lefebvre G, Dieux-Coeslier A, Mezel A, Herbaux B, Boutry N. Imaging features of lower limb malformations above the foot. Diagn Interv Imaging. 2015 Sep;96(9):901-14. doi: 10.1016/j.diii.2014.08.008. Epub 2015 Apr 24. PMID: 25920687.

Feick E, Hamilton PR, Luis M, Corbin M, Salback NM, Torres-Moreno R, Andrysek J. A pilot study examining measures of balance and mobility in children with unilateral lower-limb amputation. Prosthet Orthot Int. 2016 Feb;40(1):65-74. doi: 10.1177/0309364614560941. Epub 2014 Dec 16. PMID: 25515343.

McQuerry J, Gammon L, Carpiaux A, Talwalkar V, Iwinski H, Walker J, Jacobs CA, Muchow R. Effect of Amputation Level on Quality of Life and Subjective Function in Children. J Pediatr Orthop. 2019 Aug;39(7):e524-e530. doi: 10.1097/BPO.000000000001321. PMID: 30608302.

Ulger O, Sener G. Functional outcome after prosthetic rehabilitation of children with acquired and congenital lower limb loss. J Pediatr Orthop B. 2011 May;20(3):178-83. doi: 10.1097/BPB.0b013e3283449362. PMID: 21394040.

Fixsen JA. Major lower limb congenital shortening: a mini review. J Pediatr Orthop B. 2003 Jan;12(1):1-12. doi: 10.1097/01.bpb.0000043730.21564.92. PMID: 12488764.

Krebs DE, Edelstein JE, Thornby MA. Prosthetic management of children with limb deficiencies. Phys Ther. 1991 Dec;71(12):920-34. doi: 10.1093/ptj/71.12.920. PMID: 1946626.

Abudu A, Grimer R, Tillman R, Carter S. The use of prostheses in skeletally immature patients. Orthop Clin North Am. 2006 Jan;37(1):75-84. doi: 10.1016/j.ocl.2005.08.008. PMID: 16311113.

Hirons RR, Williams KB, Amor RF, Day HJ. The prosthetic treatment of lower limb deficiency. Prosthet Orthot Int. 1991 Aug;15(2):112-6. doi: 10.3109/03093649109164645. PMID: 1923712.

Sener G, Yiğiter K, Bayar K, Erbahçeci F. Effectiveness of prosthetic rehabilitation of children with limb deficiencies present at birth. Prosthet Orthot Int. 1999 Aug;23(2):130-4. doi: 10.3109/03093649909071624. PMID: 10493140.

Boonstra AM, Rijnders LJ, Groothoff JW, Eisma WH. Children with congenital deficiencies or acquired amputations of the lower limbs: functional aspects. Prosthet Orthot Int. 2000 Apr;24(1):19-27. doi: 10.1080/03093640008726518. PMID: 10.855435.

Downie GR. Limb deficiencies and prosthetic devices. Orthop Clin North Am. 1976 Apr;7(2):465-73. PMID: 1264434.

Bergère A, Amzallag-Bellenger E, Lefebvre G, Dieux-Coeslier A, Mezel A, Herbaux B, Boutry N. Imaging features of lower limb malformations above the foot. Diagn Interv Imaging. 2015 Sep;96(9):901-14. doi: 10.1016/j.diii.2014.08.008. Epub 2015 Apr 24. PMID: 25920687.

Di Gennaro GL, Gallone G, Martinez Vazquez EA, Marchesini Reggiani L, Racano C, Olivotto E, Stilli S, Trisolino G. Deformity progression in congenital posteromedial bowing of the tibia: a report of 44 cases. BMC Musculoskelet Disord. 2020 Jul 3;21(1):430. doi: 10.1186/s12891-020-03408-w. PMID: 32620101; PMCID: PMC7334844.

Friend L, Widmann RF. Advances in management of limb length discrepancy and lower limb deformity. Curr Opin Pediatr. 2008 Feb;20(1):46-51. doi: 10.1097/MOP.0b013e3282f35eeb. PMID: 18197038.

Fixsen JA. Major lower limb congenital shortening: a mini review. J Pediatr Orthop B. 2003 Jan;12(1):1-12. doi: 10.1097/01.bpb.0000043730.21564.92. PMID: 12488764.

Hamdy RC, Makhdom AM, Saran N, Birch J. Congenital fibular deficiency. J Am Acad Orthop Surg. 2014 Apr;22(4):246-55. doi: 10.5435/JAAOS-22-04-246. PMID: 24668354.

Özdemir M, Kavak RP, Ceylan AH, Cevval ZK. Isolated unilateral proximal focal femoral deficiency presenting in a young woman. BMJ Case Rep. 2020 Jan 21;13(1):e232714. doi: 10.1136/bcr-2019-232714. PMID: 31969407; PMCID: PMC7021155.

Research Through Innovation