

Rehabilitation Management of a Pediatric Patient with Modified Van Nes Prosthesis after Type B-III A Hip Rotationplasty: A Case Report

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ABSTRACT

Van Nes rotationplasty has gained popularity as a surgical treatment for malignant bone tumors without resorting to amputation. However, the nature of the surgery poses unique challenges in the rehabilitation management in order to optimize safety and functional mobility.

We describe a case of a 6-year-old female with Ewing's sarcoma of the right femur who underwent Van Nes Type B-III A rotationplasty. This procedure involved a complete femoral resection, with the tibia and the foot rotated by 180 degrees and the lateral tibial plateau placed into the acetabulum that is expected to remodel to form a neo-hip joint. The patient was referred to rehabilitation medicine and was eventually admitted due to impairment in mobility secondary to the surgery. The goal was to achieve K-1 level of function.

This case demonstrates that the rehabilitation management of a child with Type B-III A rotationplasty using a modified Van Nes prosthesis requires a comprehensive multidisciplinary approach.

Keywords: Van Nes rotationplasty, prosthesis, rehabilitation, neo-hip joint

INTRODUCTION

Ewing's sarcoma is the second most common malignant bone tumor, commonly affecting children and adolescents. Treatment modalities typically include a combination of chemotherapy, radiotherapy, and surgical intervention. Among the surgical options, Van Nes rotationplasty has gained popularity as an alternative to endoprosthesis replacement or traditional amputation. This procedure, when combined with a suitable prosthesis, offers functional mobility comparable to that of a transtibial amputee by preserving voluntary motion at the knee level.¹

A specific subtype, Type B-III A rotationplasty, involves placing the lateral tibial plateau into the acetabulum after rotating the tibia and foot segment by 180 degrees. Over time, this configuration is expected to remodel, allowing the knee joint to function as the new hip joint and the ankle joint to serve as the new knee joint. However, the nature of the surgery inevitably leads to muscle weakness, particularly in the hip muscles, which presents significant challenges in the rehabilitation process.²

From the standpoint of a rehabilitation medicine specialist, these challenges necessitate modifications in the rehabilitation program and prosthetic prescription. The primary goals are to optimize patient mobility, ensure the



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protection of the postoperative site, and ultimately facilitate the patient's return to independent function.

CASE PRESENTATION

Patient Information and History

A 6-year-old, Filipino student, was admitted to a tertiary government hospital in June 2022 for prosthetic training after undergoing Type B-III A hip rotationplasty. The patient was born full term to a then 32-year-old G1P0 mother via normal spontaneous vaginal delivery at a local hospital with no noted fetomaternal complications. She is an only child, living in a two-story house with her parents and grandmother. She came from a middle-income family with a significant medical history of cancer including liver, breast, cervix, and pancreas.

She achieved developmental milestones that were at par with age and was able to do independent community ambulation. However, at 5 years old, she started experiencing intermittent, right thigh pain aggravated with walking. Her mother noted a palpable, gradually enlarging, tender mass over the area (Figure 1). Persistence of symptoms prompted consult with a private physician who requested an MRI to reveal a right femoral midshaft tumor (Figure 2), and was biopsy-proven to be a round cell tumor consistent with Ewing's sarcoma. Patient then started monthly chemotherapy with alternating cycles of Doxorubicin with Cyclophosphamide and Etoposide with Ifosfamide. Serial MRI, however, revealed progression in the size of the tumor. No evidence of metastasis on other areas were found.

On October 2021, the patient underwent a Type B-III A hip rotationplasty. Postoperatively, the patient was maintained in a hip spica and later transitioned to a hip abductor brace (Figure 3). During the rehabilitation period, active movement was limited to the ankle, with no weight-bearing allowed on the neo-hip. The patient used a manual wheelchair with specific adaptations for support and comfort, relying on caregivers for mobility in the community.

Clinical Findings and Timeline

Five months post-surgery, she was cleared for fabrication of a Modified Van Nes prosthesis. This was released a month later and she had her first rehabilitation admission on the 11th of April 2022 after completing her monthly chemotherapy, for impaired mobility. The goal was to achieve K-1 level of function. It is of note, that the cost for the prosthesis was fully covered by the family.

On physical examination, the patient had a fleshy post operative site at the gluteal area, a surgically rotated right ankle that acts as a neo-knee, and her right proximal tibia acting as neo-hip (Figure 4). No significant discrepancy was noted in the level of the new, right knee joint to the contralateral side. There was no noted atrophy or swelling on all extremities, with full pulses on bilateral tibial and dorsalis pedis arteries. Spine examination, done while the patient was seated, revealed no abnormal gross curvature,



Figure 1. Pre-operative gross image showing the patient's right thigh mass.

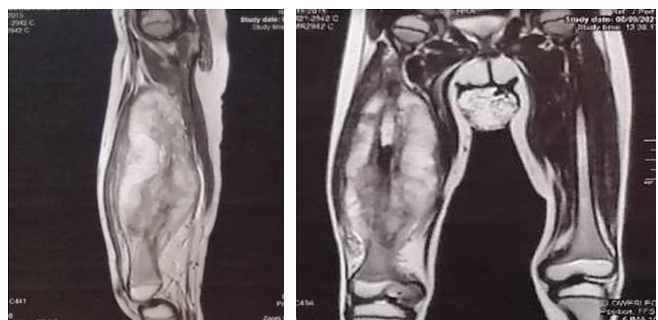


Figure 2. Pre-operative MRI of the patient's right thigh showing a femoral midshaft tumor.

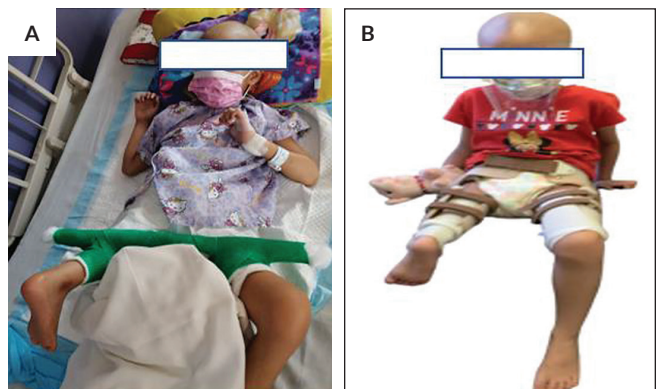


Figure 3. After undergoing surgery, (A) the patient was placed for six weeks on a hip spica to position the hips at 45 degrees flexion and 30 degrees abduction and then (B) a hip abductor brace for three more months.

or rib hump. Patient had pain-free, full range of motion on passive movement including the neo-knee and hip. She had good motor strength on the upper extremities. The unaffected, left lower extremities were graded 4/5. Motor examination for the right hip flexor, gluteus medius was 2/5, while hip abductors and adductors were 3/5. The right tibialis anterior acting as the prostheses knee joint flexor and gastrocnemius



Figure 4. Patient on admission 5 months after Van Nes Type B-III A rotationplasty showing a fleshy post-operative gluteal area, and a shortened leg with the ankle at the position of, and with the function of, an actively controlled knee. Thus, having the knee joint functioning as a neo-hip while the original ankle joint functioning as a neo-knee.

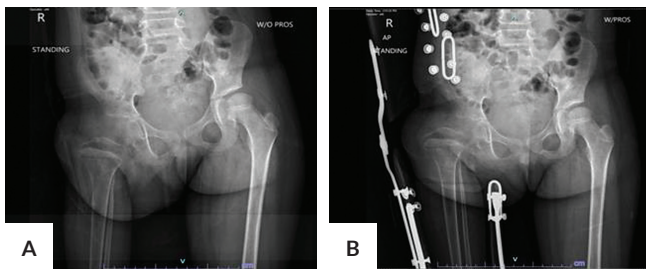


Figure 5. Standing pelvic radiograph on AP view (A) without and (B) with prosthesis. The patient's right femur was surgically resected, the tibia was rotated 180 degrees and was placed into the acetabulum. The X-ray confirmed that the lateral tibial condyle is in place and had remodeled to from a neo-hip that resembles that of a developing femoral head.

acting as the prosthesis knee joint extensor were graded 4/5. Patient had intact bilateral proprioception on bilateral extremities. Patient denied sensory deficits.

Diagnostic Assessment and Therapeutic Intervention

The design of the prosthesis for the Type B-III A rotationplasty is complex and must be highly customized to meet the unique anatomical and functional needs of the child. A key consideration in the prosthesis includes a modified Canadian socket to provide support on the neo hip joint during standing and thus, preventing subluxation. The integration of a knee mechanism that accommodates the ankle's new role and the overall alignment and biomechanics of the prosthesis were also crucial. The material is made of carbon fiber which is lightweight yet durable, and the

design allows for growth and easy adjustments. Aesthetic customization of the design, as well as choice of material in the fabrication play an important role as these can affect the acceptance and satisfaction with the use of said prosthesis.

However, upon check out of the modified Van Nes prosthesis, the modified Canadian socket and the free hip hinge required revision. X-ray on standing with and without the prosthesis were ordered to confirm the alignment of the right lateral tibial condyle as a neo-hip joint which in the case of the patient, was in place acting as the femoral head (Figure 5). The patient's rehabilitation goals were adjusted to focus on pre-prosthetic training while awaiting the prosthesis revision. The rehabilitation exercises included gluteal strengthening, neuromuscular retraining (such as figure-eight exercises), sit-to-stand training, and exercises to improve standing balance and tolerance. The patient was discharged with a home program designed to strengthen the muscles necessary for eventual ambulation with the prosthesis.

Additionally, it was noted that the patient appeared timid during interactions with paramedical staff, but a psychological evaluation revealed no signs of depression or issues with self-image.

The patient and her mother were diligent in following prescribed exercises at home, which included straight leg raises for hip flexion, assisted pelvic bridges, hip extensions, upper extremity and core resistance exercises, and sit-to-stand practices. As the patient grew stronger, she advanced to quadruped exercises and could transition from sitting to standing with minimal support, even without her prosthesis.

On June 5, the patient was re-admitted for intensive rehabilitation focusing on prosthetic ambulation using the Van Nes prosthesis with a modified Canadian socket (Figure 6). The therapy concentrated on fitting the prosthesis correctly and included training on donning and doffing it to avoid discomfort. The first week emphasized stability while standing (Figure 7) and strengthening various muscle groups through functional activities in occupational therapy and neuromuscular retraining in physical therapy. Psychological support through mother and child counseling was also done.

The rehabilitation progressed to ambulation with a pediatric walker, obstacle courses, and ramp navigation to enhance balance and coordination. After two weeks, the patient reached a K1 functional level, achieving safe and basic mobility, with emphasis on using assistive devices and making home modifications to ensure safety and improve her quality of life.

Follow ups and Outcomes

The patient was scheduled for a follow up after two weeks with plans for future re-admissions and gait analysis. However, one week post discharge, the patient experienced pain over the contralateral left thigh causing limitation and eventual discontinuation in using the prosthesis.

This case illustrates the challenges and complexities involved in the management of a child with Van Nes prosthesis



Figure 6. The patient's Van Nes prosthesis modified with a Canadian socket in order to support the Neo hip and prevent subluxation.



Figure 7. Pictures of patient standing within parallel bars while wearing the prosthesis.

with a modified Canadian socket, particularly in the context of severe and recurrent conditions like Ewing's sarcoma.

The Prosthesis Evaluation Questionnaire (PEQ) scores highlight some critical insights:

- ****Ambulation (19.60) and Utility (48.13)**** scores are significantly low, indicating difficulties the patient faced in using the prosthesis effectively. This could be due to the strength of the rotated limb, the weight of the prosthesis, and the patient's lack of acclimation to its use.
- ****Frustration (85), Perceived Response (94.30), and Social Burden (91.50)**** scores suggest that while the patient was emotionally and socially affected by the challenges, they perceived a reasonable level of support and were managing their frustrations relatively well.

- ****Residual Limb Health (79.50) and Well-being (90)**** scores are relatively higher, suggesting the patient was generally in good spirits and had no significant issues with the residual limb health during the limited time they used the prosthesis.

Unfortunately, the recurrence of Ewing's sarcoma, as evidenced by the pelvic X-ray findings, ultimately led to the patient's decline despite further aggressive treatment.

This case emphasizes the importance of ongoing monitoring, tailored prosthetic support, and the need for a multidisciplinary approach in managing complex cancer cases with prosthetic use.

Scale	Issues
Ambulation	General walking, close spaces, up stairs, down stairs, up steep hills, down steep hills, sidewalks and streets, slippery surfaces
Residual Limb Health	Sweat, smell, swelling, rash, ingrown hairs, pimples, sores
Utility	Socket fit, weight, comfort while standing, comfort while sitting, balance, energy, feel (of the surface), donning
Appearance	Look of prosthesis, damage to clothes, damage of cover, shoe choice, clothes choice
Sounds	Frequency of sound, bothersomeness of sound
Frustration	Frequency of frustration, most frustration
Perceived Response	Avoid strangers reactions, partner's response, relationship affected, family member response, other family member response
Social Burden	Burden to partner, social activity hindered, care-giving
Quality of Life /Well Being	Well-being since amputation, quality of life

Figure 8. The nine domain scales of the Prosthesis Evaluation Questionnaire (PEQ).

DISCUSSION

Ewing's sarcoma, a common primary bone cancer in children and adolescents, is characterized by the EWS-FLI1 fusion gene and commonly affects the femur, presenting with localized pain, swelling, and a soft tissue mass. Imaging typically reveals an ill-defined osteolytic lesion with moth-eaten bone destruction, and an onion skin or sunburst periosteal reaction.¹

The Van Nes rotationplasty is a surgical option for resecting malignant femoral tumors, originally used for proximal femoral focal deficiency. In Type B-III A rotationplasty, the femur is resected and the tibia is rotated 180 degrees, effectively turning the ankle into a functional knee joint.² This technique generally results in fewer complications, better outcomes, and improved gait.³ Studies and case reports, including one involving a 14-month-old patient, indicate good functional results and successful independent walking with a Van Nes prosthesis.⁴

The Prosthesis Evaluation Questionnaire (PEQ) is a 54-item self-report tool designed to assess functional outcomes in prosthetics. It includes nine domain scales and additional questions on satisfaction and prosthetic care, and has been validated for various settings (Figure 8).⁵

The patient's modified Van Nes prosthesis for a Type B-III A rotationplasty was specifically designed with a modified Canadian socket to support the right neo-hip during its remodeling phase and to prevent subluxation. The rehabilitation program was tailored to address the unique needs of the patient, focusing on neuromuscular retraining of the neo-hip and ankle joints, effective use of the modified Van Nes prosthesis with the modified Canadian socket, and providing psychological support and counseling.

A strength of this study was the individualized rehabilitation management of child with a Type B-III A rotationplasty in achieving a K1 level of functional using a modified Van Nes prosthesis with a modified Canadian socket. The PEQ was utilized as baseline functional outcome

measure during the two weeks of use of the modified van nes prosthesis that encompasses several prosthesis functions and took into account personal views and experiences of the patient, in this case, the mother.

The combined efforts of the multidisciplinary rehabilitation team enabled the patient to achieve a K1-level function, which included limited, assisted ambulation after two weeks of in-patient rehabilitation. This achievement highlighted the importance of a well-coordinated rehabilitation program in enhancing the functional outcomes for patients with complex prosthetic needs.

However, the patient's progress was complicated by tumor recurrence on the contralateral thigh, which necessitated the discontinuation of prosthesis use. This unfortunate development underscores the challenges and uncertainties that can arise in the long-term management of patients with significant medical and surgical histories, and the need for ongoing monitoring and adaptability in treatment plans.

Patient Perspective

The gratitude expressed by the mother, despite the difficulties and the loss, underscores the positive impact the rehabilitation and prosthesis had on her daughter's quality of life. The dedication of the case report to them is a meaningful tribute to their strength and the collective efforts of everyone involved in her care.

CONCLUSION

This case demonstrates that the rehabilitation management of a child with Type B-III A rotationplasty using a modified Van Nes prosthesis requires a comprehensive multidisciplinary approach. Key elements include neuromuscular retraining, tailored prosthesis design, material selection, psychosocial support, and regular monitoring. The prosthesis design should be simple, stable, and durable to achieve a K1-level of function, focusing on safe and functional basic mobility. Additionally, the Prosthesis Evaluation

Questionnaire (PEQ) proved to be a valuable tool for baseline evaluation, providing critical insights into the child's experience with the prosthesis and serving as an effective outcome measurement tool.

Ethical Considerations

Permission for the publication of case report was granted from the mother. The authors guarantee utmost confidentiality.

Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

All authors declared no conflicts of interest.

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REFERENCES

1. Burchill SA. Ewing's sarcoma: diagnostic, prognostic, and therapeutic implications of molecular abnormalities. *J Clin Pathol.* 2003 Feb; 56(2):96-102. doi: 10.1136/jcp.56.2.96. PMID: 12560386; PMCID: PMC1769883.
2. Gupta SK, Alassaf N, Harrop AR, Kiefer GN. Principles of rotationplasty. *J Am Acad Orthop Surg.* 2012 Oct;20(10):657-67. doi: 10.5435/JAAOS-20-10-657. PMID: 23027695.
3. Hillmann A, Rosenbaum D, Gosheger G, Hoffmann C, Rödl R, Winkelmann W. Rotationplasty type B IIIa according to Winkelmann: electromyography and gait analysis. *Clin Orthop Relat Res.* 2001 Mar;(384):224-31. doi: 10.1097/00003086-200103000-00026. PMID: 11249169.
4. Bhamra JS, Abdul-Jabar HB, McKenna D, Ng Man Sun S, Gillott, E, Pollock R. Van Nes rotationplasty as a treatment method for Ewing's sarcoma in a 14-month-old. *Int J Surg Case Rep.* 2013;4(10): 893-7. doi: 10.1016/j.ijscr.2013.07.027. PMID: 23978532; PMCID: PMC3785944.
5. Boone D, Coleman KL. Use of the Prosthesis Evaluation Questionnaire (PEQ). *JPO Journal of Prosthetics and Orthotics* 2006 Jan: 18(6): 68-79 doi: 10.1097/00008526-200601001-00008