Successful First Gait of a Child With Hip-Disarticulation Prosthesis

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Abstract

An infant boy underwent hip disarticulation for infantile fibrosarcoma immediately after birth. His rehabilitation began when he was 4 months old and involved training with his left (residual) leg. He could stand with support at 12 months. His initial prosthesis fitting was performed at the age of 13 months. He could stand and walk with support at 15 months of age and could walk with no additional support and go up and down stairs at 2 years. A single-axis prosthetic knee joint was introduced at the age of 2 years 3 months. His first gait using a hip prosthesis was successful, and his prosthesis was replaced at appropriate intervals with no major problems. The authors believe that the key to achieving a successful prosthetic gait in children is good communication among the medical team, which should comprise an orthopedic doctor, rehabilitation doctor, nurse, physical therapist, prosthetist/orthotist, and the patient's parents.

Keywords

hip-disarticulation prosthesis, infant, sarcoma

Text

Introduction

A malignant tumor is one cause of amputation in children. Infantile fibrosarcoma (IFS) is a soft tissue tumor occurring at an incidence of five per million infants in the pediatric age group.[1] IFS simulates classic adult fibrosarcoma histologically; however, it has a more favorable prognosis than its adult counterpart, with a mortality rate of <5% and a recurrence rate of 5% to 50%. Nearly all cases of IFS occur in the first year of life, and 36% to 80% are congenital. Treatment of IFS is primarily surgical with wide local excision. Neoadjuvant chemotherapy has been used in the last few decades to prevent extensive surgery. However, some cases of IFS may behave more aggressively, and limb salvage is not feasible because of multiple recurrences or extensive infiltration. The authors encountered a patient with IFS who required hip disarticulation immediately after birth because the tumor grew rapidly within a few days and his general condition was worsening. Nearly all children with lower limb deficiency are fitted with a prosthesis. The present patient underwent an initial prosthesis fitting that was based on his achievement of developmental milestones and an early rehabilitation intervention to introduce a single-axis prosthetic knee joint.

This study conforms to all CARE guidelines and reports the required information accordingly (see Supplementary Checklist).

Case report

An infant boy was born at 36 weeks' gestation weighing 2105 g and was the second-born of identical twins. At birth he had a 5 × 5 × 3 cm mass in his right calf, suspicious for hemangioma. He was referred to

the authors' hospital on day 12 of life because of rapid enlargement of the mass. Physical examination revealed a large, hard, elastic, non-mobile mass in his lower extremity. The overlying skin was tense and purple in color. Laboratory examination on day 11 of life showed hypercalcemia with a corrected calcium level of 11.8 mg/dL. Magnetic resonance imaging on day 17 of life showed a huge, nonspecific mass involving the neurovascular bundle and bones in his leg (Figure 1). An open biopsy was performed, and the diagnosis of spindle cell sarcoma was made. The histologic classification was not defined. A computed tomography scan of the chest and abdomen showed no evidence of distal metastases.

Because of the tumor's rapid daily growth and the increase in the serum calcium concentration to 16 mg/dL, emergency hip disarticulation was performed on day 18 of life. Upon dissection, the tumor measured 11 × 8 × 7 cm and had infiltrated the neurovascular bundles and bones around the knee. Histopathologic examination revealed a lobulated mass infiltrating the soft tissue. The mass was composed of sheets of round, ovoid, and spindle cells with necrosis, calcification, and inflammation. The tumor cells showed little pleomorphism and prominent mitotic activity. Immunohistochemical stains were negative for desmin, MyoD1, smooth-muscle actin, and CD34 and focally positive for cytokeratin, s-100, and CD99. Fluorescence in situ hybridization analysis revealed ETV6-NTRK3 gene fusion. Based on these findings, the final diagnosis was IFS. Microscopically complete resection was achieved.

Postoperatively, the patient progressed favorably, and his serum calcium concentration immediately normalized. He did not appear to have developmental delay, and he began his rehabilitation in the outpatient clinic at the age of 4 months. A physical therapist began by stretching the soft tissue of his trunk and

supporting his rolling-over motion. He could sit up on his own at 8 months of age, and sitting-posture training and supported-standing training were introduced with his left residual leg, focusing on preventing pelvic inclination and scoliosis. At 12 months of age, he could stand on his left leg while holding onto something.

The patient's first prosthesis fitting was performed at the age of 13 months, and a Canadian hip-disarticulation prosthesis without a knee joint was created (Figure 2a). The patient began prosthetic training, and at his monthly medical check, his parents received guidance in home training from the physical therapist. By 15 months of age, the patient could wear the prosthesis all day and was able to stand and walk while holding onto something. By 2 years of age, he could walk without support and was able to go up and down stairs. His artificial leg length and socket size were adjusted to accommodate growth. A single-axis prosthetic knee joint was introduced at 2 years 3 months of age (Figure 2b). His parents were eager to give him a flexible knee joint on adapting to his community life. Furthermore, the need for a flexible knee joint was increased in this case because of the Japanese custom of sitting on the floor. He performed his walking training only indoors. Although his prosthetic knee joint sometimes gave way, he sustained no serious injuries. He gradually adjusted to the prosthesis and could jump or kick a ball at 2 years 10 months of age. The patient was followed up every month for 2 years and then every 2 to 3 months for the next 2 years, and the fit and function of his prosthesis was checked at the outpatient clinic. His parents could communicate directly with the prosthetist/orthotist or his home doctor at any time. At the time of this writing, the patient was 4 years of age and had no evidence of recurrence or metastasis.

Discussion

Limb loss in children is caused by congenital disorders or amputation resulting from trauma, malignancy, infection, or other pathology. A survey on amputation from 2001 to 2005 by Ohmine et al.[2] in Kitakyushu City, Japan revealed that the annual incidence of overall amputation was 6.9 per 100,000 population, while that for lower limb amputation was 5.8 per 100,000 population. The age distribution was 0.2 per 100,000 population per year (3.4% of all ages) in individuals aged 0 to 17 years. The level of amputation was hip disarticulation in 1.7% of all lower limbs. The causes of amputation in individuals aged 0 to 17 years included congenital malformation (66.7%), malignant neoplasm (16.7%), and injuries and infection (8.3%). According to this survey, hip disarticulation for malignancy in an infant, such as the current patient, is extremely rare. IFS is a rare soft tissue tumor in infants younger than 1 year and has an estimated incidence of five new cases per million infants.[1] IFS simulates classic adult fibrosarcoma histologically, but it has a more favorable prognosis than its adult counterpart; more than 80% of patients are usually cured. The histologic diagnosis of IFS is sometimes difficult to achieve. Other differential diagnoses may also be suggested in the context of a rapidly progressive tumor, such as hemangioma, hemangiopericytoma, or myofibromatosis. Molecular biology may be useful because the presence of a specific transcript, ETV6-NTRK3, is most often associated with IFS. Although surgery should still remain the mainstay of treatment of IFS, chemotherapy has been reported to be fairly effective. An alkylating agent-free and anthracycline-free regimen is usually effective and should be chosen at first-line chemotherapy for inoperable tumors.[3] In the current patient, a spindle cell sarcoma was suggested by open biopsy, but the precise diagnosis was not

defined. Moreover, the patient was exposed to life-threatening risk in the daily disease progression; thus, emergency amputation could not be avoided.

Boonstra et al.[4] examined the functional aspects of limb loss in 88 children aged 1 to 18 years (congenital deficiency, n = 64; acquired amputation, n = 24). In 46 children with congenital lower limb deficiencies, the first prosthesis was fitted at an average age of 15 ± 5 months (range, 9–30 months). Children using prostheses in the congenital deficiencies group (n = 59) could walk at a mean age of 20 ± 6 months (range, 11–39 months). The children using prostheses with a free knee (n = 8) were fitted with first articulated knees at a mean age of 37 ± 11 months (range, 20–54 months). The first prosthesis fitting for lower extremity limb deficiency should be based on a toddler's achievement of developmental milestones. The age at which the child begins to pull to stand is the typical point for fitting because equal leg length is considered essential for normal development and independent ambulation. Clinicians are recommended to be more proactive about early prosthesis fitting for young patients.[4-6]

Prostheses are replaced more frequently for children than adults, particularly in infancy and preadolescence. Because younger children may refuse to use a prosthesis that feels uncomfortable, these children and their parents will need to visit an orthopedic or rehabilitation medicine clinic for a prosthesis that is too short, that is dysfunctional because of wear and tear, or that is ill-fitting as well as for skin problems after prosthesis fitting. The authors believe that parents' active participation is important for successful prosthetic fitting. The current patient was introduced to his first single-axis prosthetic knee joint early because he had already used the prosthesis without a knee joint well and his parents were eager to

give him a flexible knee joint to facilitate adapting to his community life. Cultural factors, such as the Japanese lifestyle in the present case, may also affect prosthesis prescription. A flexible knee joint is needed for Japanese patients who often sit on the floor. Although the optimal timing to introduce an articulated knee joint remains controversial, the authors believe that clinicians should meet the demands of the patient and family positively. In the present case, the patient's rehabilitation and instructions to his parents on proper home training were started as soon as possible after surgery. After the introduction of the patient's hip prosthesis, his parents were instructed to check the fit of the prosthesis to prevent skin problems and pain due to tightness of the socket. His prosthesis was replaced at appropriate intervals without major problems. His socket was replaced five times from the initial fitting to the age of 4 years. Adjustments in length and alignment were made more frequently to prevent the patient from walking with excessive lateral trunk bending. The patient's prosthesis will need to be replaced every 1 to 2 years, or more frequently, as he continues to grow. Further follow-up is required to provide rehabilitation with his well-fitted prosthesis for adaptation to school and social activities.

In conclusion, first gait using a hip prosthesis was successfully provided to an infant with a lower limb deficiency immediately after birth. The authors believe that the key to successful prosthetic gait in children is good communication among the medical team, which should comprise an orthopedic doctor, rehabilitation doctor, nurse, physical therapist, prosthetist/orthotist, and the patient's parents. It is particularly important that parents are able to communicate directly with the prosthetist/orthotist without frequent clinic visits.

Text

Informed consent

Written informed consent was obtained from all subjects for publication of this case report and accompanying images. A copy of the written consent is available for review upon requests.

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Figure Legends

Figure 1. On MRI, the mass has heterogenously hyperintense on coronal view (a) and axial view (b) of T2-weighted images, and it involves the neurovascular bundle and bones in his leg.

Figure 2. Photographs of the patient showing (a) initial hip disarticulation prosthesis consisting of the 7E8 modular hip joint, 2R40=2-foot adapter, and ottobock 1S30 SACH foot (Otto Bock HealthCare GmbH, Duderstadt, Germany), and (b) hip disarticulation prosthesis consisting of the ottobock 7E8 hip joint with the ottobock modular 3R38 single-axis knee joint (Otto Bock HealthCare GmbH). The foot system consists of the ottobock C1305 SACH adapter (Otto Bock HealthCare GmbH) and Lapoc C1100 VIP foot (Imasen Engineering Corp., Kakamigahara, Japan).