FLEX CEUs

Rotationplasty

Amputation and rotationplasty in children with limb deficiencies: current concepts

Abstract
Purpose Amputations and fitting surgery have a long history in children with limb deficiencies. With the current developments in limb reconstruction and new techniques in prosthetics, the indications for amputation and fitting surgery might have shifted, but still have a very important role in creating high functional performance, optimal participation and quality of life. The purpose of this current concepts article is to give an overview of the indications, dilemmas and technical considerations in the decision-making for amputation and fitting surgery. A special part of this overview is dedicated to the indications, variations and outcomes in rotationplasties.
Methods The article is based on the experience of a multidisciplinary reconstruction team for children with complex limb deficiencies, as well as research of the literature on the various aspects that cover this multidisciplinary topic.
Results For those children with a more severe limb deficiency, reconstruction is not always feasible for every patient. In those cases, amputation with prosthetic fitting can lead to a good result. Outcomes in quality of life and function do not significantly differ from the children that had reconstruction. For children with a postaxial deficiency with a femur that is too short for lengthening, and with a stable ankle and foot with good function, rotationplasty offers the best functional outcome. However, the decision-making between the different options will depend on different individual factors.
Conclusions Amputations and rotationplasties combined with optimal prosthesis fitting in children with more severe limb deficiencies may lead to excellent short- and long-term results. An experienced multidisciplinary team for children with complex limb deficiencies should guide the patient and parents in the decision-making between the different options without or with prosthesis.

Keywords Limb deformity · Limb deficiency · Postaxial hypoplasia · Syme amputation · Rotationplasty

✉ Ralph Sakkers
r.sakkers@umcutrecht.nl

[1] Department of Orthopaedic Surgery, Wilhelmina Children's Hospital, University Medical Center Utrecht, P.O. Box 85090, 3508 AB Utrecht, The Netherlands

[2] Brain Centre Rudolf Magnus and Centre of Excellence for Rehabilitation Medicine, De Hoogstraat Rehabilitation, University Medical Center Utrecht, Utrecht, The Netherlands

Introduction

Limb deformities in childhood can be congenital, as in children with pre- or postaxial hypoplasia [tibial deficiency, fibular deficiency, proximal focal femoral deficiency (PFFD) and ray defects of the foot], or acquired, as in children with progressive deformities due to sepsis or trauma involving growth plates or tumours and sequelae of the resection of tumours. In some of these children with limb deformities, the dilemma of reconstruction versus amputation will arise. Indications for reconstruction versus amputation will not only depend on the technical possibilities and the prognosis on functional outcome, but also on the wishes of the patient and the parents, the amount of time to invest in youth, the perceived quality of life during youth and in adulthood, and the cultural differences in acceptance of cosmetic appearance. A special type of functional reconstruction through a rotationplasty is another possibility in a specific group of children, but this

highly functional reconstruction will not always be acceptable from a cosmetic point of view in the patient groups with different cultural backgrounds.

This article will focus on the different aspects that play a role in the decision-making for amputations as well as the anatomical requirements (weight-bearing, stump length) for adequate prosthetic fitting at the different ages in children with congenital limb deficiencies. Indications for rotationplasties will also be discussed with and without an anomaly of the hip joint.

Primary goals in reconstruction surgery of the lower limb

The primary goal in reconstruction of a lower limb is to achieve an optimal functional outcome for each individual patient. At the end of growth, the pelvis should be horizontal in stance, the axis deformations minimalised, with the patient standing on his/her own feet or with the prosthesis on. Comfortable sitting, preferably with the knees with or without prosthesis at the same distance from the hip, compatible with most chairs should be aimed for. A good endurance in walking activities, by creating a stable weight-bearing lower extremity, and active participation in sports are other important goals. Last but not least, cosmetic appearance should be taken into account.

The dilemmas in reconstruction versus amputation

The configuration, stability and range of motion of the ankle and foot joints are the main determinants for adequate weight-bearing on the foot. The amount of rays in a foot is of lesser importance. For example, a plantigrade one-ray foot with a stable ankle joint and a minor leg length discrepancy can have a good weight-bearing capacity and a better functional outcome than a five-ray unstable valgus foot with a synostosis of talus and calcaneus in a parallel position and a complete fibular deficiency with a leg length discrepancy of more than 50%. On the other hand, equal leg length with a partial transverse deficiency of the forefoot and a stable ankle joint can result in adequate weight-bearing without shoes, but can raise such fitting problems for shoes that fitting surgery by amputation is asked for by the patient for optimal prosthesis fitting in combination with regular shoes. The advanced dynamic properties of prosthetic feet in contrast to the aplastic own foot could also contribute to the wish for an amputation.

The leg length discrepancy is another important item in decision-making. In general, leg length discrepancies in one bone larger than 30% are considered too much for reconstruction by lengthening of bone and soft tissues, especially when combined with significant joint abnormalities [1]. With modern techniques, like the newer reconstructions for the ankle, knee and hip joint, and the possibility for a more patient friendly intramedullary bone lengthening from a certain age [2, 3], the indications for lengthening are expanded. If stable joints are successfully created, larger leg length discrepancies sometimes require up to four lengthening procedures during youth starting from around the age of 2 years and might be repeated every 3–6 years [1, 4]. With a healing index ranging between 1.2 and 2 months/cm [2, 4, 5] and a lengthening of between 3 and 8 cm per lengthening of a bone per period of lengthening [1, 3, 4], the lengthening procedures might require a significant amount of time and effort by both the patient and the parents, and may interfere with the social life of the patient in his/her important childhood and adolescent years. Dealing with these interventions including dealing with pain might be too much of a burden for certain families. In our opinion, proper screening and education of both the child and the parents is necessary. The advantages and disadvantages of the different choices (amputation, reconstruction and lengthening, or prosthetic fitting without surgery) should be elicited in terms of the amount of time and effort to invest, the physical and psychological impact of the proposed procedure and the expected impact on quality of life during childhood and the quality of life in the long run. The role of other families who went through the same procedures is of utmost importance [6]. Costs of repeated surgical procedures are expensive, but in the long run, it is probably less expensive than lifelong prosthetic provisions [7]. However, functional outcome in youth and adulthood as well as a personal satisfactory cosmetic outcome in youth and adulthood are the major important determinants. In the scarce literature on long-term outcomes of patients with significant limb deformities that were lengthened with reconstruction and those patients that had a primary amputation, there was no significant difference in outcome in both performance as well as quality of life [8–12]. Limitations of these studies are heterogeneity of the groups. On the other hand, studies and institutions that report improved outcomes of newer surgical techniques lack a current objective comparison with amputation groups. Although surgical techniques have improved, prosthetic techniques have also improved in such a way that even in top-level sports, the difference is small [13, 14]. The current Olympic record (2012) for 100 m running is 9.63 s held by Usain Bolt versus 10.9 s for the "Blade Runner" Jonnie Peacock at the Paralympics (which equals 8th place in the 2012 Olympic final). The current Olympic record (2012) for the high jump is 2.38 m by Ivan Ukhov versus the record (2012) of 2.12 m by Maciej Lepiato. The current world record for the long jump is 8.95

m by Mike Powell (1991) versus 8.40 m for the "Blade Jumper" Markus Rehm (2015).

With all the different dilemmas in decision-making, the final choice remains an individual one, especially in those patients in the grey zone of indication for reconstruction or amputation [15]. This choice will be dependent on the individual's important parameters for his/her quality of life. These parameters are influenced by previous experiences, personal beliefs and adjustment capacities, and will correlate strongly to the psychological well-being, self-esteem and social circumstances of the patient and their parents [16, 17]. Therefore, a multidisciplinary treatment team, with sufficient knowledge of all important factors (physical, psychological, social) that play a role, should preferably guide the decision-making in order to get a final individualised and optimal short- and long-term outcome [18]. For a complete education, adequate photographic and video material is necessary, and contacts with individual patients and parents to share experiences, and contacts with the patient organisation should be offered. These contacts are usually much appreciated and useful for the parents to get a better idea of the functional outcomes and cosmetic appearance by seeing real children with different solutions.

In case of a choice for a primary amputation of the foot and alignment of the tibia, the preferred timing is before walking age in most cases, i.e. around the age of 1 year [19]. Sometimes, especially in cases in which the leg length discrepancy is predominantly in the femoral part and the quality of the ankle joint is good, the decision for amputation or rotationplasty will be postponed to a later age, depending on the other choices made for reconstruction.

Technical considerations in amputations and fitting surgery

One of the issues to address in amputation and fitting surgery is the choice between weight-bearing and non-weight-bearing solutions. If possible, a weight-bearing level of amputation is the first choice. Advantages of a weight-bearing stump are a less aggressive fitting of the prosthesis, distal loading through the retained heel pad, which seems to give more stability and less complications such as bony overgrowth of the bony end with possible ulceration of the skin [7, 19, 20]. In childhood, there is a possibility to stand on the stump without prosthesis, which can be an advantage in, for example, the shower or the swimming pool. Despite the progressive leg length discrepancy during growth, the weight-bearing capacities are an advantage with transfers or walking short distances in the house.

The most often used weight-bearing level of amputation below the knee is the Syme and the Boyd amputations [7, 20]. Both procedures are, in fact, ankle exarticulations that do not disturb the epiphysis. Syme described his technique of exarticulation of the foot in 1842, in which he covered the end of the tibia and fibula with the strong skin of the heel pad. The main problems described with Syme's technique are possible posterior migration of the heel pad, heel pad slough and fitting problems. However, in general, these issues are reported as not significant [9]. The Boyd amputation, described in 1939, is also a disarticulation through the ankle, but in this procedure, the posterior part of the calcaneus is preserved and fused to the distal tibia [21, 22]. The heel pad remains attached to the posterodistal part of the calcaneus, preventing heel pad migration after solid fusion of the posterior part of the calcaneus to the distal tibia. From a personal experience, a Syme amputation gives a more natural placement of the heel pad in those children that have a severe equinus hind foot with a low height of the talus calcaneus (synostosis) bone in fibular deficiency, where the heel pad just fits over the end of the tibia with or without fibula. On the other hand, an exarticulation of a foot with a normal height of the hind foot at a later age can benefit from the extra length of the part of the calcaneus distal from the tibia for a natural placement of the heel pad.

When a transtibial amputation cannot be avoided, osseous overgrowth is a well-known problem. Different techniques have been developed to prevent an open marrow space at the end of the bone, with various degrees of success [23, 24]. Especially techniques that provide an osteocartilaginous ending to the end of the bone, such as, for example, transplanting a fibular head or a metatarsal head in the end of the tibia seem to decrease the percentage of overgrowth from 50% to around 0–10% [24]. Planning on optimal stump length at adult age including the calculation of remaining growth in the growth plates at the timing of reconstructions and fitting surgery is important. The minimum stump length for adequate prosthesis fitting in below-knee amputations is 10–15 cm to preserve enough leverage for power and control of the prosthesis [25, 26]. On the other hand, the leg length difference after amputation might be too small to be able to build in a more advanced prosthetic foot in adolescence and adulthood. The optimal leg length difference is currently between 12 cm (e.g. Vari-Flex Junior) and 18–30 cm (sports foot like Cheetah Xplore) for these more dynamic prosthetic feet that are believed to have several advantages for the consumer [13, 25].

In transtibial amputees for example, there seems to be a slight trend towards a greater stride length when walking with the Flex-Foot in comparison to the SACH foot [25, 27, 28]. When walking speed was increased or when subjects were walking on a decline or incline treadmill, the energy cost was lower with an energy-storing foot than

Fig. 1 Typical example of a Boyd amputation for optimal fitting in a below-knee prosthesis

with the SACH foot [25, 29]. Most prosthetic companies have a special Syme-type prosthetic foot for adults that need a 5–6 cm average build height. Newer materials such as glass fibre composites in the RUSH® foot promise to have more dynamic properties and durability even in the low build-height feet for active kids and adults.

Thus, if possible, a weight-bearing level of amputation like a Syme or Boyd amputation, combined with an epiphysiodesis to create the optimal leg length difference for the more advanced prosthetic feet seems to be the best option. This option can only be created during the growth period in childhood (Figs. 1 and 2).

Technical and functional possibilities with rotationplasties

Indications for rotationplasties in patients with postaxial hypoplasia are usually seen in patients where PFFD is the dominating factor and the ankle and the foot have a (near) normal anatomy and function. Mild PFFD may be treated with guided growth, alignment surgery and/or lengthening techniques. However, in femurs with length discrepancies of over 30–50%, lengthening might not be possible and one has to find other ways for a better functional outcome. An extra important goal is comfortable sitting, preferably with the knees at the same distance from the hip, and a length compatible with most chairs.

Borggreve is known to be the first surgeon to create a functional knee joint by excising the original knee with part of the distal femur and proximal lower leg with sparing of the neurovascular structures. With subsequent 180° rotation of the distal part of the lower leg with a well-functioning ankle

Fig. 2 Five-year old boy with a congenital knee disarticulation fitted with a blade for running

and foot and fusing this part to the proximal part of the femur, he created a new 'knee' joint with the ankle for fitting into a below-knee prosthesis without the foot sticking out. This procedure was performed in a 12-year-old boy who had a stiff knee from tuberculosis [30]. In 1937, Van Nes performed rotationplasty in a patient who had PFFD and this procedure became popular in the 1940s and 1950s. The procedure became known as the Van Nes rotationplasty [6, 8, 19, 31–34].

The Van Nes rotationplasty is a creative solution for PFFD with high functional outcomes and high outcomes of quality of life, despite reported reduced symmetry in stance and reduced endpoint and maximum excursions [8]. Modifications have been made to prevent the derotation of rotated segment by de-attaching all muscle tendon units and re-attaching these 180° opposite of the joint and to address different levels of resection of bone, including a tibial rotationplasty [34–36].

Winkelmann made a modified classification of rotationplasties on the basis of which part of the leg had to be resected in oncology [37, 38]. The procedure in AI and AII lesions resulted in a femur with a normal hip joint and an ankle as the knee joint. The procedure in the BI and BII lesions have a 180° rotated knee fused to the pelvis after resection of the largest part of the femur without or with

part of the pelvis. The knee is fused at the level of the hip joint and points downward to the floor, acting as a hip joint with one axis. Flexion of the 'hip' occurs with flexion of the knee and extension of the 'hip' with extension of the knee at the level of the pelvis. With lesion BIIIa, the total femur is removed, the rest of the leg is rotated 180° and the tibial plateau is placed in the acetabulum. In children below the age of 10 years, the (original) lateral part of the tibia plateau can even be remodelled to a new ball joint in the acetabulum.

Projecting this classification and procedures on patients with PFFD, the more severe PFFD patients in which lengthening procedures will not produce an adequate length of the femur can be divided into two groups: those patients with a functional hip joint and those patients without a functional hip joint.

In general, the group of patients with PFFD with a functional hip joint might need a procedure to optimise the hip apart from other reconstructive procedures. Part of these patients will need an acetabular and or femoral correction osteotomy for a better stability and long-term outcome of the hip. The next question is if and how to create a functional length from hip to knee. Some patients accept a short femur and difference in knee height and use a lengthening prosthesis for walking. Other patients prefer a knee arthrodesis and the foot at the level of the knee to fit into a prosthesis in which the foot can actively control the prosthesis with a hinge at the level of the opposite knee of the normal leg [39]. This solution has the disadvantage that the foot is sticking out at the level of the knee with problems of clothing and cosmetic issues. Therefore, some patients choose for a Syme or Boyd amputation at the level of the opposite knee joint. This gives a much better cosmetic appearance, however at the cost of a functional joint at the level of the knee and, therefore, a lower functionality of the total leg and a higher energy expenditure during walking and other activities [39]. The best functional procedure that can be offered is the rotationplasty comparable with Winkelmann AI and Winkelmann AII result in combination with a specific rotationplasty prosthesis [19, 33, 34, 36, 38].

For the group of patients without a functional hip joint, the main goal is also to create a functional upper part of the leg that can be fitted with a below-knee prosthesis. Several solutions have been created. This group can be divided into a group with a knee joint and a group without a knee joint.

In the group with a knee joint and a short distal femur, the patient can be fitted with a prosthesis with ischial support. Sometimes, it is useful to perform an early epiphysiodesis of the distal femur to keep this part as short as possible for better fitting of the prosthesis during growth. Some of these patients will choose for a knee fusion with or without a Syme or Boyd amputation.

Also in this group, different types of rotationplasties have been described. One method is femoropelvic fusion with the distal femur parallel with the ground, resulting in a knee acting as a one-axis hip joint with 90° flexion (full extension of the knee at the pelvic level) [40]. Epiphysiodesis of the short distal part of the femur was needed in some of the cases to prevent growing of the distal part of the femur in the anterior direction and some patients had a rotationplasty in the lower leg or a Syme amputation [41]. The method with resection, rotationplasty and femoropelvic fusion, in which the short distal femur is rotated 180° and fused to the pelvis with the knee pointing downwards to the floor and the knee acting as a hip joint with one axis, similar to the procedure for Winkelmann BI, can be used in PFFD without a hip joint. The method similar to the procedure for Winkelmann BIIIa in order to create a ball joint at the articulation of the original lateral part of the tibia plateau and the acetabulum can be used in young patients [42]. Prerequisites are a functional ankle and foot that can be fitted in a rotationplasty prosthesis.

In all procedures, the heel should end a few centimetres short compared to the level of the contralateral patella with sitting in order to have equal thigh length on both sides with sitting with the prosthesis on the leg. Although all

Fig. 3 Nine-year-old girl with rotationplasty (procedure as for Winkelmann A1) for postaxial hypoplasia with dominant proximal focal femoral deficiency (PFFD) component. Length is calculated on the remaining growth of the *left* leg

Fig. 4 Optimal function with a rotationplasty prosthesis on a rip stick after further growth of the left femur at the age of 14 years, with the 180° rotated ankle joint at a level a few centimetres above the contralateral knee joint

reports on quality of life scales in patients with a Van Nes rotationplasty and modifications generally do not show any difference with a control population [43], the biggest controversy for this procedure for some patients is the cosmetic appearance without prosthesis, which makes this procedure not acceptable for every suitable candidate (Figs. 3, 4 and 5).

Conclusion

Amputation and fitting surgery in children with severe limb deficiencies can lead to very functional and satisfactory results. Even in those children in whom reconstruction is possible, an individual cost–benefit ratio should be made. When the investment in time and effort for reconstruction is very high, patients and parents might also choose for amputation and fitting surgery. Functional outcome and quality of life might be equal or even better in individual cases. A rotationplasty can be the best long-term functional option in patients with a postaxial deficiency with a functional ankle and foot, and a femur that is too short for lengthening. The biggest controversy in this highly functional option is the cosmetic appearance without prosthesis. In general, multiple issues regarding surgery, prosthetic

Fig. 5 Optimal function with a rotationplasty prosthesis on a rip stick after further growth of the left femur at the age of 14 years, with the 180° rotated ankle joint at a level a few centimetres above the contralateral knee joint

prescription and rehabilitation arise in decision-making in patients with more severe limb deficiencies. The overall purpose is to create a high functional performance, optimal participation and quality of life, and outcome is influenced by multiple (physical, psychosocial and social) factors. The incidence of limb deficiencies is low and, therefore, knowledge should be centred. The impact on childhood and adult life can be high, and the choices to be made should be guided by an experienced multidisciplinary team that is also able to evaluate new innovative techniques in both surgery and prosthetic design in a broad reference spectrum.

Compliance with ethical standards

Ethical approval This article does not contain any studies with human participants performed by any of the authors.

Rotationplasty with Vascular Reconstruction for Prosthetic Knee Joint Infection

Rotationplasty is used most often as a function-preserving salvage procedure after resection of sarcomas of the lower extremity; however, it is also used after infection of prosthetic knee joints. Conventional vascular management during rotationplasty is to preserve and coil major vessels, but recently, transection and reanastomosis of the major vessels has been widely performed. However, there has been little discussion regarding the optimal vascular management of rotationplasty after infection of prosthetic knee joints because rotationplasty is rarely performed for this indication. We reviewed four patients who had undergone resection of osteosarcomas of the femur, placement of a prosthetic knee joint, and rotationplasty with vascular reconstruction from 2010 to 2013. The mean interval between prosthetic joint replacement and rotationplasty was 10.4 years and the mean interval between the diagnosis of prosthesis infection and rotationplasty was 7.9 years. Rotationplasty was successful in all patients; however, in one patient, arterial thrombosis developed and necessitated urgent surgical removal and arterial reconstruction. All patients were able to walk independently with a prosthetic limb after rehabilitation. Although there is no consensus regarding the most appropriate method of vascular management during rotationplasty for revision of infected prosthetic joints, vascular transection and reanastomosis is a useful option.

1. Introduction

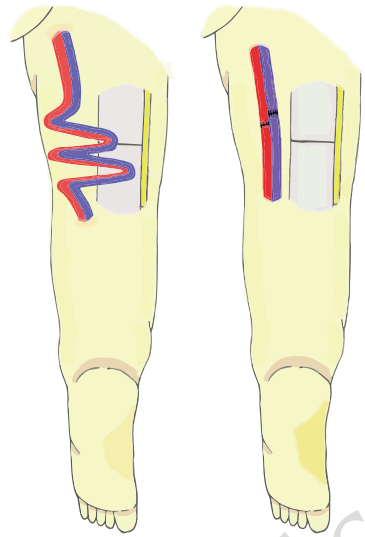
Rotationplasty, in which the distal leg and foot are rotated axially 180 degrees and grafted to the femur to create a functional joint replacing the knee, has been frequently performed as an alternative to above-the-knee amputation for skeletally immature patients after resection of sarcomas of the lower extremity [1–7]. However, rotationplasty can also be performed as a function-preserving salvage procedure for adult patients with infected knee prostheses [8–11].

Conventional vascular management during rotationplasty is to preserve major vessels and coil them medially with the distal leg, but recently, the utility of transection and reanastomosis of the major vessels has been widely reported (Figure 1) [1, 2, 12–14]. However, no consensus has been reached regarding the optimal method of vascular management of rotationplasty after the resection of sarcomas [1, 12, 13]. Furthermore, there has been little discussion regarding the vascular management of rotationplasty for revision of infected prosthetic knee joints [1].

In the present study we reviewed four patients who had undergone rotationplasty with vascular reconstruction for revision of tumor prosthetic knee joints after long-term infections and summarize the relevant literature to guide management of such cases.

2. Patients and Methods

We reviewed four patients who underwent Van Nes rotationplasty, which allows continuation of longitudinal bone growth, for the revision of infected knee prostheses, at the National Cancer Center Hospital from 2010 to 2013. The patients were three men and one woman, with a



(a) (b)

Figure 1: (a) Preservation and coiling of the major vessels, (b) transection and reanastomosis of the major vessels.

Table 1: Patient characteristics and operative details.

Patient	Age (years), sex	Interval from joint replacement to rotationplasty (years)	Interval from infection to rotationplasty (years)	Operation time (hours)	Donor vessels	Recipient vessels	Perioperative complication	Follow-up after rotationplasty (months)
1	17, male	5.0	2.7	9.3	Femoral artery/vein	Popliteal artery/vein	None	44
2	22, male	12.0	6.0	13.2	Femoral artery Femoral vein Branch of femoral vein	Posterior tibial artery Peroneal vein Posterior tibial vein	None	39
3	45, female	18.6	18.3	16.4	Femoral artery/vein	Popliteal artery/vein	Arterial thrombosis	12
4	20, male	5.8	4.6	13.2	Femoral artery/vein	Popliteal artery/vein	None	10

mean age of 26.0 years (range, 17–45 years), who had received tumor prosthetic knee joint replacement after the resection of osteosarcomas of the femur. The mean interval between prosthetic joint replacement and rotationplasty was 10.4 years (range, 5.0–18.6 years) and the mean interval between the diagnosis of prosthesis infection and rotationplasty was 7.9 years (range, 2.7–18.3 years). All patients selected rotationplasty because of the pain and decreased range of motion of the infected prosthetic knee joints. The mean follow-up period after rotationplasty was 26.3 months (range, 10–44 months) and no patient was lost during follow-up.

The rotationplasty procedure was performed as follows. First, circumferential incisions were made in the skin of the distal thigh and proximal leg. Next, osteotomies were proximally and distally performed at the levels of the joint prostheses in the femur and the tibia, respectively. The sciatic nerve was then dissected and preserved along the entire length of the resection. Major vessels were excised en bloc with soft tissue around the knee joint, and the remnant leg and foot were externally and axially rotated 180 degrees. Femorotibial osteosynthesis was performed before vascular reconstruction and achieved via intramedullary nails or plate fixation. Vascular reconstruction was then performed under an operating microscope, and the order of arterial and venous anastomosis was determined in accordance with the intraoperative setting of vessels. Finally, the wound was closed in layers.

3. Results

The characteristics of the patients and operative details are summarized in Table 1. The mean operation time was 13.0 hours (range, 9.3–16.4 h). In three patients, the femoral artery and vein were anastomosed to the popliteal artery and vein, respectively. In one patient (patient 2), the femoral artery was anastomosed to the posterior tibial artery, and the femoral vein and its branch were anastomosed to the peroneal vein and the posterior tibial vein, respectively, because the vessels had been transected distally at the bifurcation of the popliteal vessels.

Although the postoperative course was uneventful in three patients, arterial compromise developed on the first postoperative day in one patient (patient 3). To restore blood flow, urgent surgical removal of a thrombus and arterial reconstruction with a vein graft were performed. On the other hand, no patients developed venous compromise after surgery.

Palsy of the sciatic nerve did not develop in any patient. In addition, all patients started rehabilitation approximately one week after rotationplasty; all were able to walk independently with a prosthetic limb during the last follow-up.

4. Representative Case

Patient 2 was a 22-year-old man who presented with an infected prosthetic knee joint. When he was 11 years old, he underwent surgical excision with a wide margin and neoadjuvant chemotherapy for an osteosarcoma of the left femur and then received a prosthetic knee joint. However, an infection developed in the prosthetic joint 6 years later. Because of the pain and dysfunction of the affected limb, rotationplasty was performed at the patient's request.

During the operation, blood flow from the stump of the femoral artery was poor because of a thrombus of unknown cause in the proximal femoral artery; therefore, vascular reconstruction was performed after removal of the thrombus to resect the thrombosed femoral artery and perform end-to-end anastomosis.

The postoperative course was uneventful, and the patient started walking independently with a lower limb prosthesis 4 months after surgery. At 25 years of age, he is able to ride a bicycle and participate in sports while wearing the prosthesis (Figure 2).

5. Discussion

The most devastating complication after rotationplasty is vascular compromise of the rotated limb, which can result in eventual above-the-knee amputation or hip disarticulation. Although reported rates of vascular compromise after conventional rotationplasty range from 3.7% to 15.4% [1, 2, 12], the rate only after rotationplasty for revision of infected prosthesis has never been reported. In the conventional method of Van Nes rotationplasty, the femoropopliteal artery and vein are preserved with the sciatic nerve, the neurovascular structures are coiled, and the distal limb is rotated axially 180° and reattached to the proximal stump. However, these redundant and coiled vessels are susceptible to kinking and collapse, which sometimes result in vascular compromise of the distal limb. Therefore, some authors have suggested that, during rotationplasty, transection and reanastomosis of the vessels be performed [4, 14]. Although such a procedure requires technical expertise, loops of redundant vessels are removed, and the risk of kinking is reduced.

Regarding the optimal method of vascular management of rotationplasty after the resection of primary sarcomas, no consensus has been reached [12, 13]. However, vascular transection and reanastomosis would be the preferred vascular management method for rotationplasty performed for revision of infected prostheses. First, the presence of severe scar formation around the major vessels, caused by previous operations and long-term infection, could make dissection extremely difficult. En bloc resection that includes the major vessels may shorten the operation time and reduce the risk of inadvertent vessel injury. Second, the risk of vascular kinking is higher in cases of rotationplasty for revision of infected prostheses than in cases of rotationplasty for primary sarcomas when the vessels are preserved. Owing to scar formation, the major vessels lose elasticity and are more likely to become kinked [15].

The main disadvantage of transection and reanastomosis of the vessels is the risk of anastomotic failure. Although reported rates of vascular compromise after rotationplasty with vascular reconstruction range from 11.5% to 15.0% [2, 13, 16], the rate only after rotationplasty for revision of infected prosthesis has never been reported. In the present study, arterial thrombosis developed in one case; this is because of vascular kinking due to mismatch of the lengths between the remnant femur and the reconstructed vessels. In this case, we performed vascular anastomosis before osteosynthesis because the distal limb began to show signs of ischemia during resection. As a result, trimming of the artery was considered inappropriate and the artery became kinked. Therefore, we believe that osteosynthesis should be completed before vascular anastomosis unless acute revascularization is necessary.

We reviewed our four cases who had undergone rotationplasty with vascular reconstruction for revision of tumor prosthetic knee joints after long-term infections. Despite the small number of patients, the results of our cases are of interest because rotationplasty is rarely indicated for revision of infected prosthetic joints. Although there is little discussion regarding the most appropriate method of vascular management during rotationplasty for this indication, we consider that vascular transection and reanastomosis is better than vascular preservation.

6. Conclusions

Transection and reanastomosis of the major vessels is a useful option of vascular management during rotationplasty for long-term infection of knee prosthesis.

FIGURE 2: (a) Preoperative status with decreased range of motion due to infection of the prosthetic knee joint, (b) vascular reconstruction after rotation of the distal leg, (c) appearance 3-year after rotationplasty, (d) X-ray finding 3-year after rotationplasty.

Long-term functional outcome and quality of life following rotationplasty for treatment of malignant tumors

Abstract

Background: Malignant bone tumors of the lower extremity are more frequently found in children and adolescents than in adults. Modern treatment regimens led to high limb salvage rates and offer the choice between endoprosthetic replacement and rotationplasty in many cases. Rotationplasty has proven to be an effective, highly functional option in short- and mid-term studies. Aim of this study was to assess long-term results regarding quality of life and functionality after rotationplasty and to compare the obtained results to a representative healthy German sample cohort.

Methods: In total 12 patients who underwent rotationplasty between 1991 and 2001 were enrolled in this study. After physical examination, they were evaluated regarding health related quality of life, functional outcome and psychosocial status. While quality of life was mainly assessed using the SF-36 (The Short Form (36) Health Survey v2), functional outcome was measured using the musculoskeletal tumor society score (MSTS) as well as the Tegner activity level scale.

Results: Average age at the time of surgery was 19 ± 10 year. and 32 ± 11 year. at the time of follow up. Mean follow-up was 14 ± 9 years. The SF-36 scores accounted for 80.4 ± 15.7 regarding physical functioning, for 78.1 ± 24.1 regarding the physical role functioning, for 74.1 ± 17.6 regarding bodily pain and for 71.8 ± 26.1 regarding general health. SF-36 score for vitality was 75.0 ± 12.8, for social functioning 98.9 ± 3.6, 88.2 ± 23.9 for emotional role functioning and 89.6 ± 10.1 for the mental health. Comparison to a representative German sample cohort revealed significantly higher patient's scores for vitality, social functioning and mental health ($p < 0.05$). The overall MSTS resulted in an average of 64 ± 12 % and the Tegner activity level scale accounted for 4.1 ± 0.6 pts.

Conclusions: The presented long-term results indicate that rotationplasty provides a high quality of life. Patients are satisfied with a good functional outcome regarding activities of daily life and even sports.

Keywords: Rotationplasty, Bone tumor, Sarcoma, Quality of life, Function, Long-term

Background

Sarcomas account for approximately 1 % of all adult cancers [1]. In this context soft tissue sarcomas typically occur in middle aged and older adults [1], whereas malignant bone tumors, *i.e.,* osteosarcoma and Ewing's sarcoma are more frequent in children and adolescents [2, 3]. Progress in modern treatment regimens including neoadjuvant and adjuvant therapy has markedly improved the overall survival rates over the last decades [4]. Therefore, analysis of long-term outcome after malignant bone tumor therapy gains more and more importance. Moreover only a minor percentage of patients require primary amputation due to the efforts of limb preserving surgical procedures [3, 5–9]. Nowadays endoprosthetic knee replacement is associated with good functional, cosmetic and psychological outcomes, resulting in significantly better walking efficiency and musculoskeletal tumor society scores (MSTS) in comparison to major amputation or arthrodesis [10–12].

In contrast rotationplasty has been proven to be associated with equivalent functional outcomes, but better quality of life as well as less limitations during daily activities and less pain in the short- and mid-term outcome [13–17]. However, especially the cosmetic result is not as appealing as for traditional limb sparing alternatives. In this context only a small number of studies investigated the health-related quality of life (HRQL) and the long-term outcome after rotationplasty for the treatment of malignant bone tumors [18–20]. Therefore the aim of this study was to assess long-term results following rotationplasty with regard to HRQL, functional performance and psychosocial aspects and to compare theses results to a representative healthy German sample cohort [21].

Methods

Ethical approval for this project was granted by the local ethics committee (Klinikum rechts der Isar, Medical Faculty, reference no. 4092/11). Written informed consent was obtained from each patient prior to enrolment in the study.

Patients

All patients who had been treated by rotationplasty for malignant bone or soft tissue tumors of the lower extremity at our academic musculoskeletal tumor center (MSTC) between May 1991 and June 2001 were identified from our database.

Questionnaires

Before assessing the questionnaires all patients were clinically examined by an expert orthopedic surgeon (GG).

The Short Form Health Survey (SF-36v2) was used for the assessment of HRQL [22]. The SF-36v2 is a questionnaire assessing the individual health of patients as well as disease-related distress by eight scaled dimensions. Each scale ranges from zero (poor) to 100 (excellent). The eight dimensions are *vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning* and *mental health* [22].

The functional outcome was evaluated using the musculoskeletal tumor society score (MSTS) [23]. The MSTS evaluates the functional outcome of tumor patients after completed therapy. It consists of six components for the lower limb: pain, function, emotional acceptance, need for walking aids, walking and gait. The maximum (best) score for each item is 5 (range from 0 – 5). The values of each of the six components are added and divided by the maximum possible number of pts. (30). The percentage value is obtained by multiplying the calculated point value by 100.

In addition the Tegner activity level scale [24] was assessed. The Tegner activity level scale ranges from zero to ten. While an activity level of zero means sick leave or disability because of knee problems, a level of five means that the patients are able to perform heavy labor and recreational sports twice a week and a level of ten means that the person pertains to the national elite in competitive sports [25].

Statistics

Statistical analysis was performed using Sigma Stat 3.1 software (Systat Inc, Chicago, Illinois, USA. Unless otherwise stated the data is given in means (arithmetic mean) ± standard deviation (SD). Welch's *t*-test (two-sample unpooled *t*-test for unequal variances) was calculated for comparing SF-36v2 results of our patients to a representative healthy German sample cohort [21] using QuickCalcs software (GraphPad Software Inc, La Jolla, California, USA). A p-value < 0.05 was considered statistically significant.

Results

Patients

Overall 23 patients underwent rotationplasty between 1991 and 2001. Nine patients deceased due to the malignant disease, two patients were lost for follow-up. Summarizing, twelve patients (7 male, 5 female) with a median age at the time of treatment of 19 ± 10 year. and a median age of 33 ± 11 year. at follow up were enrolled. The mean follow-up was 14 ± 9 years. The diagnoses consisted of osteosarcoma ($n = 9$), chondrosarcoma ($n = 2$) and one synovial sarcoma (for patient's details see Table 1). Patients, suffering from osteosarcoma were treated according to the Cooperative Osteosarcoma Study Group (COSS) protocol and received multidrug chemotherapy before and after surgery [26]. The patient suffering from

Table 1 Overview of patient characteristics including diagnosis, treatment, complications and postoperative social status ($n = 12$)

Gender	
male	7
female	5
Age at surgery (yrs of age)	
Mean	19 ± 10
Median (Range)	18 (4–46)
Follow up (yrs)	
Mean	14 ± 3
Median (Range)	15 (8–18)
Diagnosis	
Osteosarcoma	9
Chondrosarcoma	2
Synovial sarcoma	1
Anatomic Site	
Distal Femur	9
Proximal Femur	3
Treatment	
Borggreve Rotationplasty	10
Winkelmann Rotationplasty	2
Winkelmann Classification	
A I	8
B I	1
B II	2
B IIIa	1
Complications	
Implant loosening	1
Impingement	1
Achilles tendon tear	1
Marital state	
Married	5
Divorced	-
Never Married	7
Living together with Partner	1
Living separated from Partner	1
Single living with parents	1
Single living alone	4
Education	
Less than compulsory	1
Compulsory	8
Postcompulsory	3
University level	-
Employment state	
Full-time job	7
Part-time job	1
Student	3
Illness retirement	1

synovial sarcoma underwent additional neoadjuvant and adjuvant chemotherapy following the consensus of our interdisciplinary tumor board.

Referring to the classification according to Winkelmann et al. in eight cases an AI type, in one case a BI, in two cases a BII and in another case a BIIIa-rotationplasty was performed [27].

Complications

One patient underwent one revision surgery once due to prolonged healing of the osteotomy and implant loosening. Another patient had to be treated for impingement of the Borggreve joint. One patient needed ten revision procedures due to traumatic Achilles tendon tear with consecutive postoperative wound infection. There were no further major complications recorded.

Health-Related Quality of Life (HRQL)

Analysis of the psychosocial outcome, measured by the SFv2-36 revealed for the subcategory physical health 80.4 ± 15.7 for the dimension *physical functioning*, 78.1 ± 24.1 for *physical role functioning*, 74.1 ± 17.6 for *bodily pain* and 71.8 ± 26.1 for *general health*.

Scores for the subcategory mental health state were 75.0 ± 12.8 for *vitality*, 98.9 ± 3.6 for *social functioning*, 88.2 ± 23.9 for *emotional role functioning* and 89.6 ± 10.1 for *mental health* (for details see Table 2). The comparison of the presented results to a representative healthy German sample cohort (mean age 49 ± 18 years) revealed significantly higher scores for the patients regarding the dimensions *vitality* ($p = 0.0243$), *social functioning* ($p = 0.0001$) and *mental health* ($p = 0.0001$) [21].

Functional outcomes

The overall MSTS resulted in an average of 64 ± 12 % with 63 ± 15.4 % for male and 66 ± 5.7 % for female patients. Analysis of the subcategories revealed 4 ± 0.8 pts. for pain, 3.3 ± 1.2 for function, 3.3 ± 1 for emotional acceptance, 2.8 ± 0.6 for walking supports, 3.3 ± 0.4 for walking distance and 2.6 ± 1 for gait (see Fig. 1). The Tegner activity level scale accounted for 4.1 ± 0.6 pts (see Fig. 2).

Ten patients reported no or occasional pain, two complained about moderate daily pain. Only five patients reported limitations during recreational activities and one patient was limited during daily activities. The walking distance of nine patients was greater than 6 blocks and within the range of 4–6 blocks for the remaining three patients. Five patients had no and seven patients some difficulties while walking on uneven terrain. In seven patients the gait pattern was not or slightly and in five patients obviously altered.

Ten patients reported, having little problems in participating in sports, one of the patients is a national

Table 2 Results of the SFv2-36 questionnaire assessing function and mental health of patients after rotationplasty of our series and a representative German sample [21]

Subjects	Physical functioning	Physical Role	Bodily pain	General health	Vitality	Social role	Emotional role	Mental health
Rotationplasty ($n = 12$)	80.4 ± 15.7	78.1 ± 24.1	74.1 ± 17.6	71.8 ± 26.7	75.0 ± 12.8	98.9 ± 3.6	88.2 ± 23.9	89.6 ± 10.1
Representative healthy German sample cohort ($n = 2043$) [21]	87.2 ± 20.4	81.8 ± 23.1	79.3 ± 25.3	64.4 ± 15.2	65.3 ± 18.3	87.5 ± 19.3	84.7 ± 22.7	72.3 ± 17.2
p	0.1635	0.6064	0.3309	0.3581	0.0243	0.0001	0.6229	0.0001

champion in handicapped swimming and the one patient with Achilles tendon tear performs no sports at all.

Regarding range of motion of the neo-knee flexion and extension corresponded to the levels of the contralateral ankle joint with a mild restriction (10°) in eleven patients and a moderate restriction 15° in one patient. In- and eversion was normal in all patients. The joint was regarded stable (varus/valgus stability) in all patients.

Four patients even had children following the rotationplasty. Only one patient is currently not able to work because his work as storekeeper requires carrying heavy loads. All but one patient, who was not sure, would choose a rotationplasty as treatment option again, facing the choice between amputation, endoprosthetic knee replacement or rotationplasty.

Discussion

The objective of this retrospective study was to evaluate the long-term outcome as well as health-related quality of life (HRQL) in patients who underwent rotationplasty for treatment of a malignant bone or soft tissue tumour. At a mean follow-up of 14 ± 9 years. HRQL, assessed by the Short Form Health Survey (SF-36v2) revealed good to excellent scores for each dimension. Interestingly the scores for vitality, social functioning and mental health were significantly higher compared to a representative healthy German sample cohort [21]. Functional long-term outcome measured by the musculoskeletal tumor society score (MSTS) and the Tegner activity level scale also revealed a good functional outcome with 64 ± 12 % and 4.1 ± 0.6 pts., respectively. All but one patient, who was not sure, would choose rotationplasty as therapy option again.

Rotationplasty implies significant shortening of the leg and rotation of the foot 180° around the vertical axis [28]. Therefore the former ankle adopts the role of a neo-knee with ankle dorsiflexion simulating knee flexion [29, 30]. Although the procedure allows for a function more akin to transtibial amputation due to retention of voluntary control of motion at the 'knee' level the significant cosmetic alteration implies potential future socio-psychiatric issues that might impact health and HRQL [31]. However, in our cohort we found good to excellent results regarding the subcategory physical health as well as the mental health state. These findings are in line with the work of Veenstra et al. who reported that the subdivisions daily emotional interaction, emotional support in problematic situations and social companionship are comparable to a healthy control group [32]. Consistent with previous studies, the majority of our patients presented well-adjusted in terms of social integration [33]. Though Veenstra et al. also reported almost half of the patients had negative effects of the surgery on initiating social or intimate contact, body

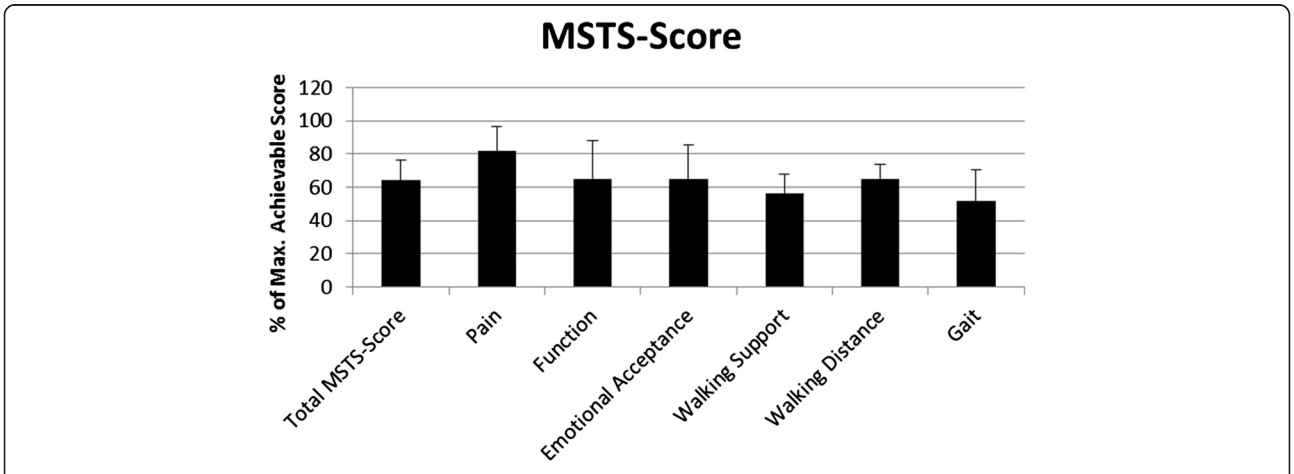


Fig. 1 Percentage of maximum MSTS-Score and sub-scores of patients after rotationplasty

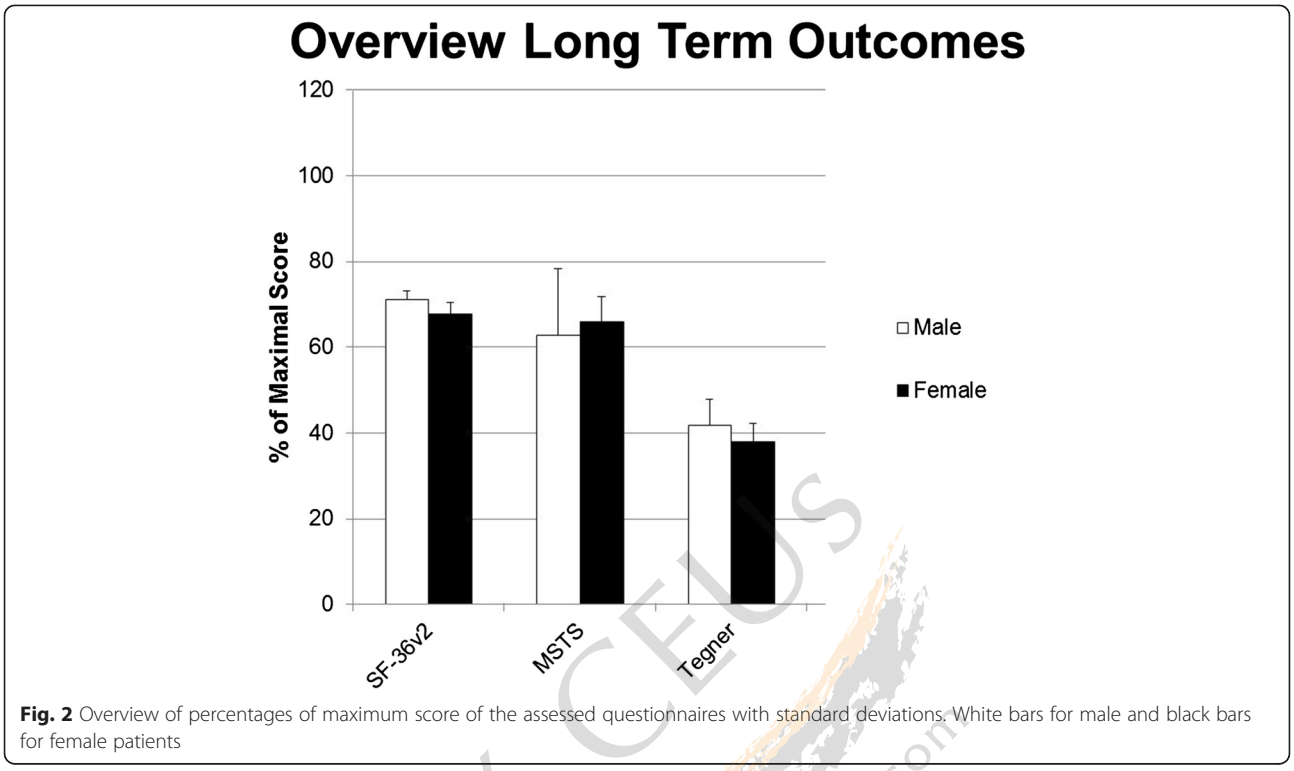


Fig. 2 Overview of percentages of maximum score of the assessed questionnaires with standard deviations. White bars for male and black bars for female patients

image [32]. In this context patients after treatment with mega-prosthesis seem to be more satisfied [32, 34]. At least patients in our study had children following the rotationplasty in four cases.

One of our unexpected findings was, that scores for vitality, social functioning and mental health were significantly higher in our series in comparison to a representative German sample [21]. At first glance this finding is hard to understand. However, in comparing our findings to the results of long-term survivors of other malignancies, our study might underline the concept of posttraumatic growth [35, 36]. In this context Sears et al. reported that 83 % of breast cancer patients felt at least one benefit in their disease [36]. These women mentioned that following cancer diagnosis and consecutive therapy they live now more intensively and consciously. Furthermore, women evaluated the social support they received during the illness as a positive aspect. This seems to be an important aspect of coping with the illness, as it has been demonstrated that social support is a significant predictor for a better long-term quality of life in breast cancer patients [36].

Of course functional outcome has a significant impact of on HRQL, as a significant reduction of HRQL was reported in patients with a MSTS <50 % [37]. In our study only one patient with an MSTS of 47 % was found to be below the 50 % threshold. This patient had sustained an Achilles tendon tear with several consecutive revision surgeries. Our other patients presented with an average MSTS of 64 ± 12.41 %. This is in line with the literature reporting MSTS between 63 and 80 % following rotationplasty [38, 39]. Comparative studies showed significantly higher MSTS-scores after endoprosthetic knee replacement than after rotationplasty [39]. Tunn et al. report MSTS-scores of 77 to 87 % after endoprosthetic limb salvage therapy of primary bone tumors in proximal tibia and distal femur, respectively [40]. However, there are also higher rates of postoperative complications reported following endoprosthetic knee replacement. In this context Warrener et al. noted complications in 42 % of patients following arthroplasty and only in 25 % following rotationplasty [39]. Aseptic loosening and mechanical failure are the most common complications [41]. Additionally, rising numbers of periprosthetic infections with multidrug resistant bacteria cause increasing concern [42]. Our patients were highly satisfied with a stable new joint offering high functionality for daily activities. The nature of the operation obviously causes difficulties with complex movements such as heel walking, jumping or walking on uneven terrain [30, 31]. However, most of our patients reported little problems in participating in sports, one of the patients is even a national champion in handicapped swimming. With an average Tegner activity level scale of 4.1 our patients felt able to perform moderately heavy work and some recreational sports. In comparison a healthy cohort achieved nearly two grades more (5.7) and was therefore able to perform heavy labor, competitive sports and recreational sports several times a week. This

seems to be in line with the work of Hillmann et al., who reported that 85 % of the patients following rotationplasty were actively participating in "high-level" sports [43].

Limitations

Several strengths and limitations of the present study have to be considered. On the one hand we focus on an extremely rare surgical entity and enrolled at least 12 patients. The average follow-up of 14 ± 9 years. is, according to the best of our knowledge one of the longest, reported in literature. Furthermore all enrolled patients were clinically seen by a doctor and fulfilled the questionnaires under advice. However, due to the rarity of rotationplasty the small number of patients limits this retrospective study. Moreover, a significant number of patients had deceased due to the disease. This might create a potential bias. Additionally the age at the time of surgery as well as the underlying diagnosis was quite heterogeneous.

Conclusions

Since limb amputation is associated with lower functional outcome, lower health related quality of life and no positive effects on survival compared to both rotationplasty and endoprosthetic reconstruction, alternative treatment options should be considered first [44–46]. Although recent studies show that modern endoprosthetic reconstructions show equal or even better functional outcomes than rotationplasty, endoprostheses bare the risk of high complication rates [38–42]. The presented long-term results indicate that rotationplasty provides a high HRQL and that patients are satisfied with a good functional outcome regarding activities of daily life. In terms of vitality, social function and mental health our study group showed even significant higher scores compared to a representative German sample in the SF-36v2 survey. In this regard we consider our work to be of distinct relevance regarding the discussion of surgical treatment plans for patients suffering from malignant tumors of the lower thigh and the choice whether to perform a rotationplasty or a endoprosthetic reconstruction.