Limb Salvage in Bone Sarcomas in Patients Younger Than Age 10. A 20-Year Experience.

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ABSTRACT

The authors present their experience over the last 20 years in limb salvage procedures of a consecutive series of 40 children un-der 10 years of age (range 2–10 years) with bone sarcomas. Nineteen were osteogenic sarcomas and 21 were Ewing sarcomas. Only one case, located in the distal phalanx of the toe, was treated by straight-forward amputation. Intercalary allografts and Cañadell's technique were used to preserve joints whenever possible, and prosthesis or osteoarticular allografts were used when the joint surface was involved. Survival rate in this series was 75%. There were four local recurrences. At the last follow-up (mean 11.2 years, range 5–19 years postop), 90% of the patients preserved their limbs. Eighty percent of the authors' results were excellent or good according to the Musculoskeletal Tumor Society Scale. Limb salvage is a real possibility even in young children with bone sarcomas. The age of the patient itself is not a contraindication for limb salvage.

KEY WORDS

Limb salvage; children; bone

INTRODUCTION

Bone sarcomas are more frequently seen in children and adolescents.[7,17–19] In the past, all these patients were treated by amputation, but the survival rates were very poor. Chemotherapy has changed this perspective in terms of survival and limb salvage possibilities. However, every institution has its own point of view regarding the possibilities of limb salvage in each case. Even within the same hospital, the criteria for limb salvage are not the same as time goes on.

From a purely oncologic point of view, in almost every case of bone sarcoma it is possible to perform a limb salvage procedure, and in experienced institutions it does not therefore compromise patient survival when compared with amputation. From a functional point of view, however, there is no agreement among institutions. One of the most controversial points is limb salvage in children.[5,21] Many believe that the best option in young children is amputation when the tumor is located in the lower limb, since it avoids growth-related problems. Several surgical techniques have been described for these cases, such as expandable prosthesis, bone lengthening, contralateral epiphysiodesis, extemporaneous lengthening by using allografts or prosthetic implants longer than the resected pieces, and so forth.[3–6,8–16,20,22,24–26] All these techniques have potential complications. Another possibility is Van Ness rotationplasty, but it is really an intercalary amputation, and it is difficult to accept in many cases due to its cosmetic appearance.[2]

We present our experience in the last 20 years in limb salvage procedures of a consecutive series of 40 young children with bone sarcomas. When possible, we preserved the growth plate near the tumor.[20,22] We used in some cases bone lengthening by external fixation and/or contralateral epiphysiodesis to correct limb length discrepancies several years after tumor surgery.[13,16,22]

METHODS

Between 1980 and 1998 we treated in our institution 506 patients with bone sarcomas.[7,17,19] Forty of them were children under 10 years of age (range 2–10 years). Table 1 shows the main data of the series. Nineteen were osteogenic sarcomas and 21 were Ewing sarcomas. The most common location was the distal femur, followed by the proximal tibia. Most of them (33 cases) were IIb lesions. Seven patients had pulmonary metastases at diagnosis. Only one case, located at the distal phalanx of the toe, was treated by straightforward amputation. Three Ewing cases were not surgically treated. Therefore, all but four cases were treated by limb salvage procedures in addition to chemotherapy and/or radiotherapy, according to our protocols (Table 2). Van Ness rotationplasty was not performed in any case.

The minimal follow-up was 5 years (except in cases of death), the maximum 19. The same medical team treated all patients, including surgeons, pediatric oncologists, and radiation oncologists. Follow-up was made every month during the first year (the year of chemotherapy), every 3 months the second year, every 6 months the third, fourth, and fifth years, and then every year. The function at last follow-up was measured according to the Musculoskeletal Tumor Society Scale (MSTS).[1] In lower extremity locations, leg length discrepancy was particularly monitored and treated when necessary. Limb

discrepancies in the upper limb did not require treatment. An inquiry was performed to assess the opinion of the patients about their surgical treatment at the last follow-up.

RESULTS

Mean follow-up for the living patients of the series was 11.4 years (range 5–19). Fourteen patients died between 6 months and 12 years after diagnosis. Four of them died of a cause independent from the disease (three due to toxicity of treatment and one due to a traffic accident 12 years after diagnosis, having no evidence of disease). Therefore, the survival rate in this series was 65% if we include those who died of an independent cause, or 75% if we do not. There were four local recurrences; three of them were treated by amputation. The other one could be removed, and a new allograft was implanted. Locations such as the proximal fibula, clavicle, or rib did not require reconstruction (five cases). For diaphyseal (and also for metaphyseal not involving the growth plate) tumors (17 cases), we used intercalary allografts. When the joint surface was resected, we employed prosthetic reconstructions (seven cases), osteoarticular allografts (five cases), or arthrodesis (one case). According to the MSTS scale, intercalary reconstruction had a better functional outcome compared with articular reconstructions (see Table 3).

Infection was present in four cases, fracture of the allograft in six cases, and nonunion in three cases. Most of these complications could be solved with further operations (see Table 1). One patient required an amputation after several operations, including lengthening, due to complications from surgery (vascular involvement and bypass thromboses).

Limb discrepancies in the upper extremity did not re-quire any surgical treatment, since the only problem is cosmetic. Depending on the possibilities in every case, lengthening was performed through the healthy or the affected bone (femur or tibia) or both. The criteria for doing a lengthening procedure included dissymmetry longer than 4 cm and at least 3 years free of disease.[13,16,22] One patient required a Judet quadricepsplasty after correction of her whole discrepancy (9 cm) by lengthening through an allograft fracture (Fig. 1). Taking advantage of some other complication, we could also perform limb lengthening in two cases. Mean lengthening was 8.9 cm. The most problematic location regarding ultimate growth was the distal femur. One patient required 32 cm of lengthening in three surgical steps.

The mean number of further major operations in the affected limb was 1.1 per patient (range 0–5). Other minor surgeries were performed, such as skin grafting, hemorrhage drainage, contralateral epiphysiodesis, and so forth. This number was higher in survivors. Therefore, the number of patients who did not require any further operation, including those who died, was 19. Only six survivors did not require a further operation. In several cases, more than one complication (eg, limb length discrepancy plus allograft fracture) could be resolved at the same surgical step. One patient who underwent amputation required a further operation for correcting a painful scar in the stump.

All but one patient said they were very happy. Even those who underwent amputation said they did not regret having tried to preserve their limbs in the first operation; at least, then, they knew that both the medical team and their parents tried to do it. The only disappointed patient had several operations after resection of a distal femur

osteosarcoma when he was 6 years old. As a result, he has very poor functionality, but he is now an adult and does not want to undergo an amputation.

DISCUSSION

This is a consecutive series of patients under 10 years of age treated by limb salvage procedures. No exclusion of patients with metastasis at diagnosis was made. A 20-year experience in a single institution is discussed. Only one straightforward amputation (in a case located at the distal phalanx of the toe) was performed. We have no experience with Van Ness rotationplasty. Ewing sarcoma is usually less frequent than osteogenic sarcoma,[7] but it is the predominant type of bone sarcoma in young children. Ewing sarcoma is the most sensitive bone sarcoma to radiotherapy. This part of treatment can increase the possibilities of local control and survival, but it also has an important influence on the functional results. In addition, radiotherapy may cause radiation-induced sarcomas. We did not see such a complication in this series.

Even in distal locations such as the ankle, it is possible to preserve a very useful limb. Some authors advise an amputation in such a location, believing that a below-knee amputation will provide better functional results. One of our patients (patient 38) is a soccer player in his village team, 15 years after reconstruction with an intercalary graft of his distal fibula. He does not remember the operation (he was 4 years old) and has never had any functional restriction (Fig. 2).

Consolidation of allografts is easier in children than in adults.[23] Only three patients required autologous bone graft supplementation to achieve consolidation. After these operations, there were no pseudarthroses.

Intercalary reconstructions had a better outcome in terms of function, lower number of complications, and so forth. This is why we perform epiphysiolysis before excision of selected cases of metaphyseal bone sarcomas in children.[20,22] This technique is used when tumor does not invade the epiphysis. We started this technique in 1984, and we continue using it in young children and also in adolescents. It allows the preservation of most of the growth plate, together with the joint. Depending on several factors, such as the type of osteosynthesis used for reconstruction, these growth plates may continue growing. In any case, functional results are better when the joint can be preserved.

The growth plates near the knee are the most important in terms of the ultimate growth of the lower limb. Chemotherapy usually inhibits growth in all growth plates, but once chemotherapy ends, the growth plates recover their potential of growing. In contrast, the damage of radiotherapy on the growth plate is permanent. Also, the age of the child is important to predict the final discrepancy: the younger the child, the longer the dissymmetry. Surgical resection of a growth plate may be necessary to remove the tumor. Reconstruction may also cause damage of another growth plate (eg, when using a knee prosthesis). Sometimes, however, the prosthesis can allow some growth if the prosthetic stem crossing the growth plate is thin enough.

In some cases, to avoid or to diminish final discrepancy, we can use a reconstruction method longer than the resected piece. However, only 2 or 3 cm can be corrected in that way because of the risk of neurovascular damage. Chemotherapy can also play a role in the possibilities of neural damage. When the expected final discrepancy is not too long,

contralateral epiphysiodesis may be the easiest way to treat the dissymmetry. A shortening osteotomy may be necessary if discrepancy is longer than 4 to 5 cm.

Several kinds of growing prosthesis have been developed in recent years.[8,11,24] The mechanisms of lengthening are now less invasive than they were before, and thus the risk of infection is now lower. The risk of fibrosis in these kinds of reconstruction is also important, and it can compromise joint motion. We have used most times fresh-frozen allografts for reconstruction, since they have better biologic integration. However, they can also have complications such as nonunion, fracture, or infection.

Bone lengthening is, in our opinion, the most physiologic way of correcting limb discrepancies in these patients.[13,16,22] We applied it when discrepancy is greater than 4 cm and the patient has been free of disease for at least 3 years. The timing of lengthening and callus formation is similar to those in other lengthening procedures because the patient is not receiving any cytostatic agents during the procedure.

Both amputation and limb salvage have similar results in terms of survival and local control. In this series survival was found to be similar to overall series of bone sarcomas, as well as the risk of local recurrence. Limb salvage may have in young children a higher number of complications than amputation, but amputation cannot be converted into a limb salvage procedure; limb salvage, however, can be converted into an amputation if it fails. Future function is the only reason for choosing amputation or limb salvage in many cases, and many times it cannot be predicted. Even in long-term follow-up studies, there are no differences in terms of economic cost between the techniques.[12]

Doctors must explain to parents the possibilities of preserving the limb in these cases. Patients and parents usually do not accept amputation, at least not as the first surgical treatment. Sometimes it may be better to amputate rather than to perform many surgeries to preserve the limb, but you cannot predict at the time of diagnosis what will happen in the future. Most of the patients come to our institution looking for a limb salvage procedure. In many cases, some other doctors had advised an amputation based on the age of the patient, the location, and the difficulties in reconstruction, and most of them are now very happy with their limbs. Fortunately, the predictions of those doctors were not fulfilled in most cases. Only four (10%) patients required a secondary amputation, while 90% preserved their limbs. This series shows that limb salvage is a real possibility even in young children with bone sarcomas. Age itself should not be the main cause for choosing an amputation in young children.

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Patient	Age/ Gender	Location	Size (cm)	Histology	Tumoral Necrosis	Surgery	Radiotherapy (rads)
1	10 M	Humerus	25	E.S.	100%***	—	4000
2	10 M	Distal femur	20	O.S.	>90%	Intercalary	—
3	10 F	Arm	12	E.S.	100%	—	4000
4	10 M	Clavicle**	15	E.S.	100%	Resection	5500
5	10 F	Proximal humerus	15	O.S.	>90%	Prosthesis	—
6	10 M	Rib	15	E.S.	>90%	Resection	4000
7	10 F	Fibula	20	E.S.	>90%	Resection	6200
8	10 M	Humerus	13	E.S.	100%	Intercalary	6200
9	10 M	Prox. tibia	12	O.S.	70%	Intercalary	—
10	10 M	Tibia	6	E.S.	100%	Intercalary	6000
11	10 F	Distal tibia	11	T.O.	20%	Intercalary	—
12	10 F	Distal femur*	12	OS.	>90%	Prosthesis	—
13	9 F	Distal femur*	21	C.O.	>90%	Intercalary	—
14	9 F	Clavicle	15	E.S.	100%	Resection	—
15	9 M	Prox. tibia	14	C.O.	40%	Osteoarticular	—
16	9 M	Pelvis	10	E.S.	100%	Intercalary	—
17	9 M	Foot phalanx	2,5	E.S.	100%	Fing. amputat	—
18	9 M	Prox. femur	18	E.S.	100%	Prosthesis	4600
19	9 F	Distal tibia	20	OS.	60%	Ankle arthrodesis	—
20	9 F	Distal femur	24	OS.	100%	Intercalary	—
21	9 F	Dist. femur*	10	OS.	>90%	Prosthesis	—
22	8 M	Distal femur*	14	OS.	>90%	Osteoarticular	——
23	8 M	Pelvis	5	E.S.	100%	Intercalary	4500
24	8 M	Distal tibia*	14	T.O.	>90%	Osteoarticular	—
25	8 M	Proximal tibia	14	E.S.	100%	Intercalary	6000
26	8 M	Distal radius	10	E.S.	100%	Autograft	5800
27	8 M	Proximal humerus	19	OS.	>90%	Prosthesis	—
28	8 M	Dist. femur	14	OS.	75%	Osteoarticular	—
29	8 M	Prox. femur	15	E.S.	100%	Prosthesis	4700
30	7 F	Prox. tibia	16	E.S.	100%	Intercalary	4600
31	7 F	Prox femur*	10,5	OS.	100%	Prosthesis	—
32	7 M	Scapula	10	E.S.	100%	—	4500
33	6 M	Distal femur	18	OS.	75%	Intercalary	—
34	6 F	Distal tibia	13	T.O.	20%	Osteoarticular	—
35	6 F	Dist. femur	12	E.S.	>90%	Intercalary	3700
36	5 M	Prox. tibia	12	OS.	100%	Intercalary	—
37	4 F	Rib	10	E.S.	100%	Resection	3000
38	4 M	Dist. fibula	8	E.S.	100%	Intercalary	—
39	3 F	Distal femur	8	OS.	>90%	Intercalary	—
40	2 M	Distal tibia	12	E.S.	20%	Intercalary	—

Table 1. (Continued)

Complications	Treatment	Length Discrepancy (cm)			Treatment of Metastasis	Oncologic Status	Follow-up (years)
		Pre	Treatment	Post			
Elbow stiffness	MUA	—	—	—	—	D.O.D.	2
Local recurrencence/ infection	Resection/ amputation	—	—	—	—	N.E.D.	9
Fracture	Osteosynthesis	—	—	—	—	N.E.D.	12
—	—	—	—	—	Bone marrow autotransplant	D.O.D.	5
—	—	—	—	—	—	D.A.C.	3.5
—	—	—	—	—	—	N.E.D.	19
Peroneal n. palsy	—	—	—	—	—	D.O.D.	1.5
—	—	—	—	—	—	D.O.D.	4
Nonunion	AGIF	—	—	—	—	N.E.D.	12
Infection	Amputation	6	Lengthening + epiphysiodesis	—	—	N.E.D.	15
—	—	—	—	—	—	N.E.D.	7
—	—	—	—	—	Thoracotomy	D.O.D.	1
Nonunion	Lengthening	10	Femur lengthening	—	Thoracotomy	N.E.D.	16
—	—	—	—	—	—	N.E.D.	7
—	—	5	Shoe supplementation	5	—	N.E.D.	6
—	—	—	—	—	—	N.E.D.	5
—	—	—	—	—	—	N.E.D.	8
Dislocation	Reduction	7	Femur lengthening	—	—	N.E.D.	16
Local recurrence	Amputation	—	—	—	—	N.E.D.	1.5
Allograft fracture	New allograft + lengthening	9	Femur lengthening	—	—	D.O.D.	11
—	—	—		—	—	D.A.C.	0.5
Allograft fracture	New allograft + arthrodesis	24	(2) Femural + (1) tibial length.	3	Thoracotomy	N.E.D.	15
—	—	—	—	—	—	N.E.D.	5

—	—	—	—	—	Thoracotomy	D.A.C.	4
Allograft fracture	New allograft	4	Femur lengthening	—	—	N.E.D.	10
—	—	—	—	—	—	D.O.D.	1
—	—	—	—	—	—	D.O.D.	1.5
Infection	New allograft	10	Femur lengthening	—	—	N.E.D.	14
Loosening	New prosthesis	10	—	10	—	N.E.D.	14
Nonunion	Autografting	8	Tibial lengthening	—	—	N.E.D.	15
—	—	8	Femur lengthening	—	(3) Thoracotomy	D.O.D.	7
—	—	—	—	—	—	D.A.C.	12
Local recurrence + infection	New allograft	35	(2) Femural + (1) tibial lengthening	3		N.E.D.	17
Allograft fracture	AGIF	—	—	—	—	N.E.D.	9
—	—	—	—	—	—	D.O.D.	3
Allograft fracture	AGIF	6	Epiphysiodesis	2	—	N.E.D.	10
Thoracic deformity	Plastic reconstruct.	—	—	—	—	N.E.D.	16
—	—	—	—	—	—	N.E.D.	16
Local recurrence	1. New allograft 2. Amputation	—	—	—	—	N.E.D.	9
—	—	6	Tibial lenghtening	—	—	N.E.D.	6

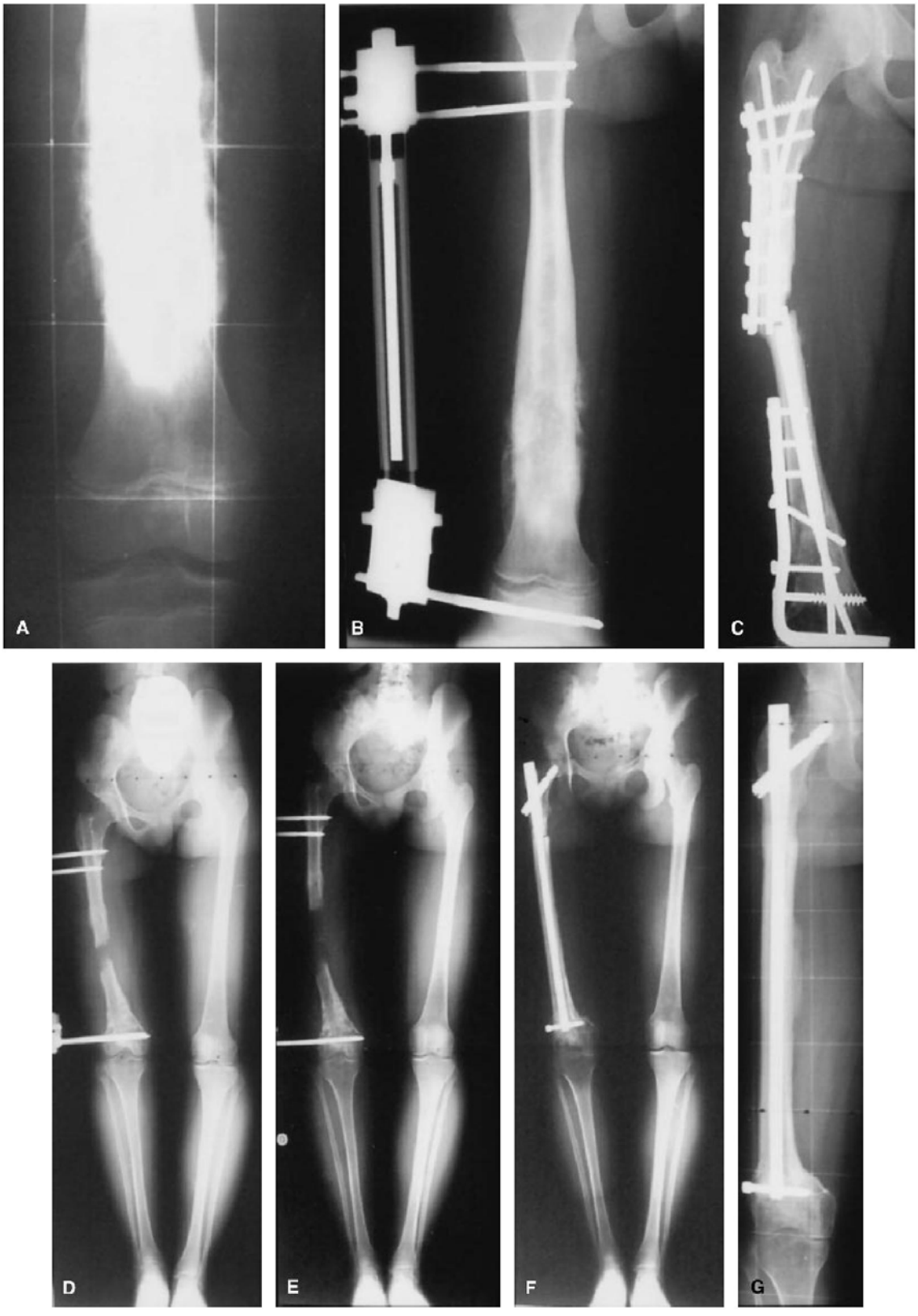
E.S., Ewing sarcoma; O.S., osteogenic sarcoma; T.O., telangiectatic osteosarcoma; C.O., chondroblastic osteosarcoma; MUA, mobilization under anesthetics; AGIF, Autologous grafting and internal fixation; D.O.D., dead of disease; N.E.D., no evidence of disease; D.A.C., dead from another cause. *Pulmonary metastasis at diagnosis; **pulmonary and bone metastasis at diagnosis; ***according to post chemo and radiotherapy multiple biopsies.

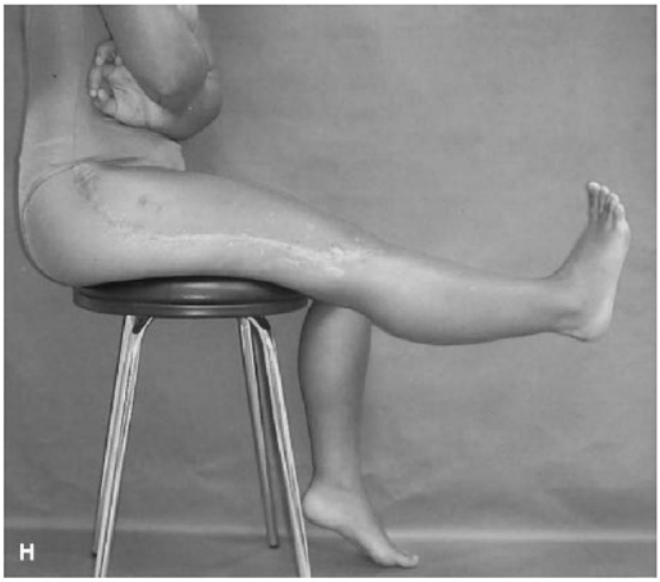
Table 2. Chemotherapeutic Protocol for Sarcomas in Pediatric Patients, University of Navarra, Pamplona, Spain
Osteosarcoma
1. Neoadjuvant chemotherapy
Cisplatinum (CDDP) 40 mg/m^2 intra-arterial (i.a.) days 1, 2, 3, 22, 23, 24, 43, 44, 45
Adriamycin (ADR) 30 mg/m^2 intravenously (i.v.) days 1, 2, 3, 22, 23, 24
2. Tumoral resection
3. Postoperative chemotherapy
• Cycle A: CDDP 40 mg/m^2 (i.a.) and ADR 30 mg/m^2 (i.v.) for 3 days
• Cycle B: (After 3 weeks)
High-dose methotrexate (HDMTX) 12 g/m^2 (i.v.) for 1 day
Cyclophosphamide 500 mg/m^2 (i.v.), actinomycin D 0.5 mg/m^2 (i.v.) and bleomycin 10 mg/m^2 (i.v.) for 1 day (within the same week as HDMTX)
• Cycle A: HDMTX 12 g/m^2 (i.v.) for 1 day
CDDP 40 mg/m^2 (i.a.) and ADR 30 mg/m^2 (i.v.) for 1 day (within the same week as HDMTX)
Alternate cycles B and A until approximately 1 year after the beginning of treatment.
Ewing Sarcoma
Cycle A
• Adriamycin (ADR) 20 mg/m^2 intravenously (i.v.) and methotrexate (MTX) 12 mg/m^2 (i.v.), days 1, 2 and 3
• Cyclophosphamide 1,200 mg/m^2 (i.v.) and vincristine 1.5 mg/m^2 on day 1
Cycle B
• Bleomycin 10 mg/m^2 (i.v.) and actinomycin D 0.5 mg/m^2 (i.v.) on days 21, 22, and 23
• Cyclophosphamide 1,200 mg/m^2 (i.v.) and vincristine 1.5 mg/m^2 (i.v.) on day 21
Notes
• Alternate cycles A and B each 21 days until completion of 9–10 months of treatment.
• In addition to chemotherapy the protocol includes local radiotherapy (in some cases) and tumor surgery.

Table 3. Functional Results in All the Cases Treated (MSTS Scale). The Two Cases Located in the Rib Were Excluded.

Site of tumor	Excellent	Good	Fair	Poor
Distal femur (9)	1	6	1	1
Proximal tibia (6)	4	0	2	0
Distal tibia (6)	4	0	2	0
Proximal humerus (5)	2	1	2	0
Proximal femur (3)	0	1	2	0
Fibula (2)	1	0	1	0
Clavicle (2)	1	1	0	0
Pelvis (Illium) (2)	2	0	0	0
Scapula (1)	1	0	0	0
Distal radius (1)	0	1	0	0
Phalanx of toe (1)	1	0	0	0

Figure 1. An 8-year-old girl diagnosed with osteosarcoma in the distal femur in 1989 (A). She was treated by epiphysiolysis, resection of the tumor, and intercalary reconstruction (B). Five years later she had an allograft fracture (C). We took advantage of this event to correct both fracture and limb discrepancy (9 cm) (D–G). She also required a Judet quadricepsplasty (H–L), but she finished growth with an excellent limb function (M–O).

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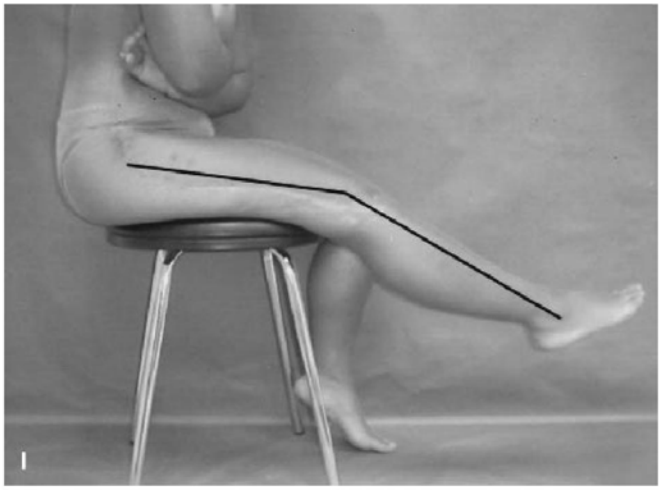
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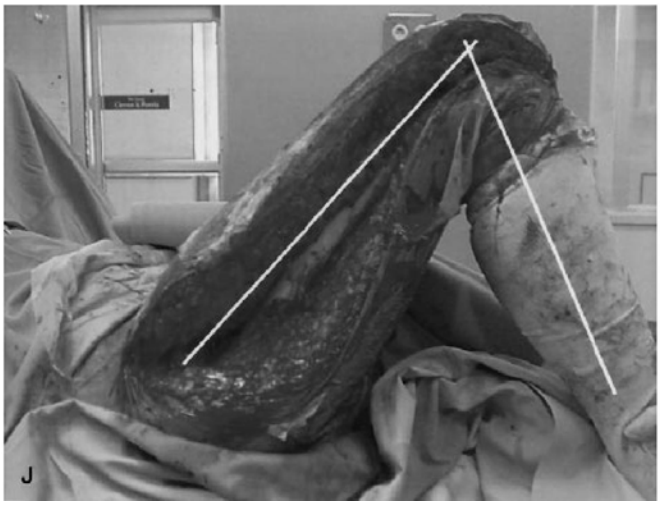
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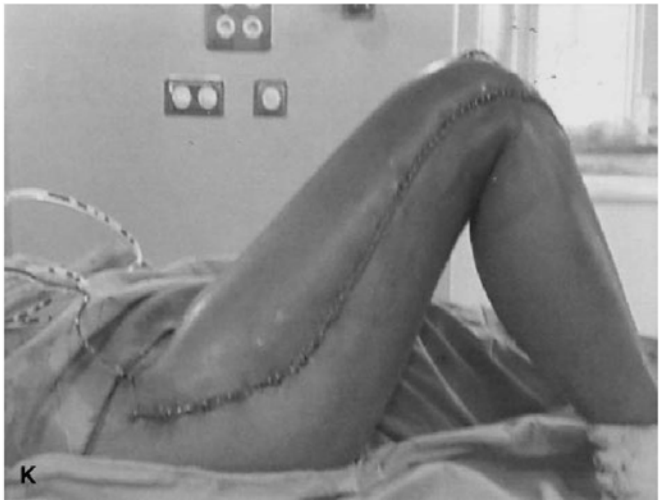
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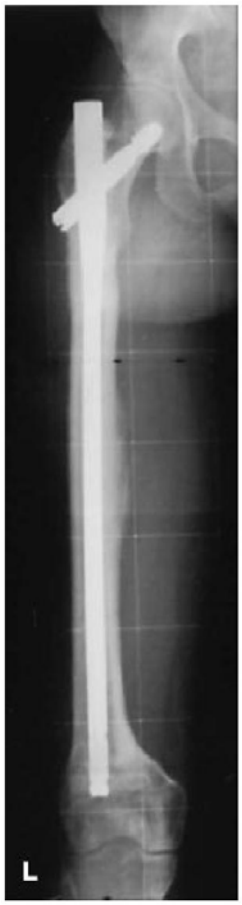
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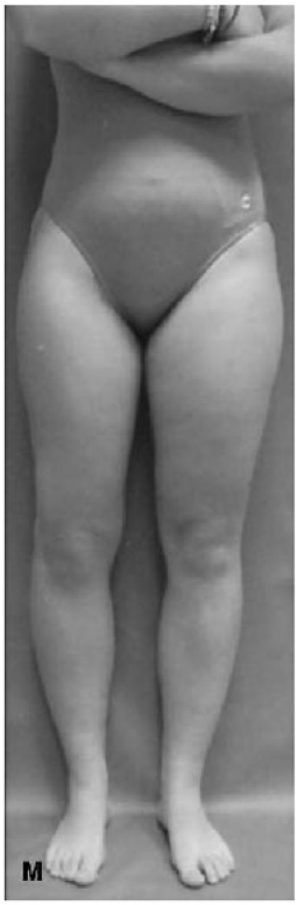
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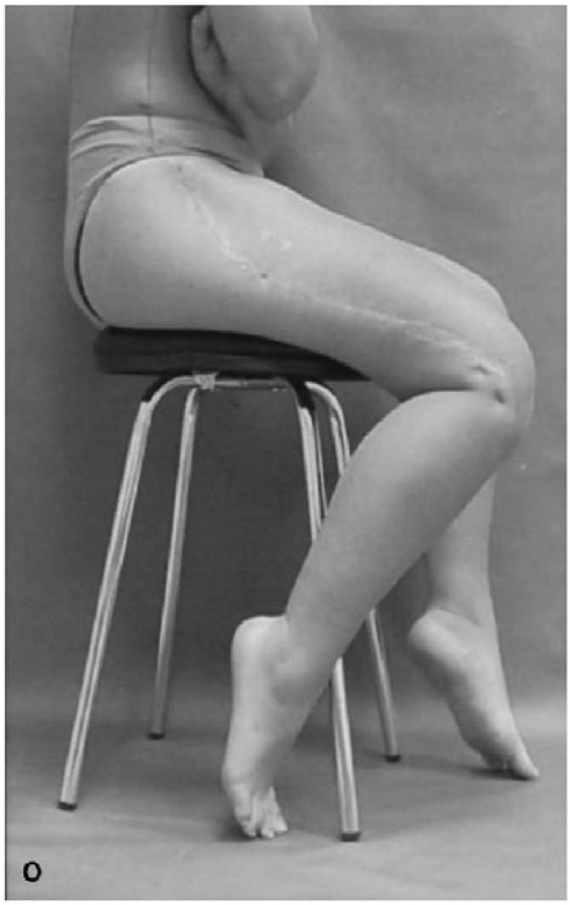
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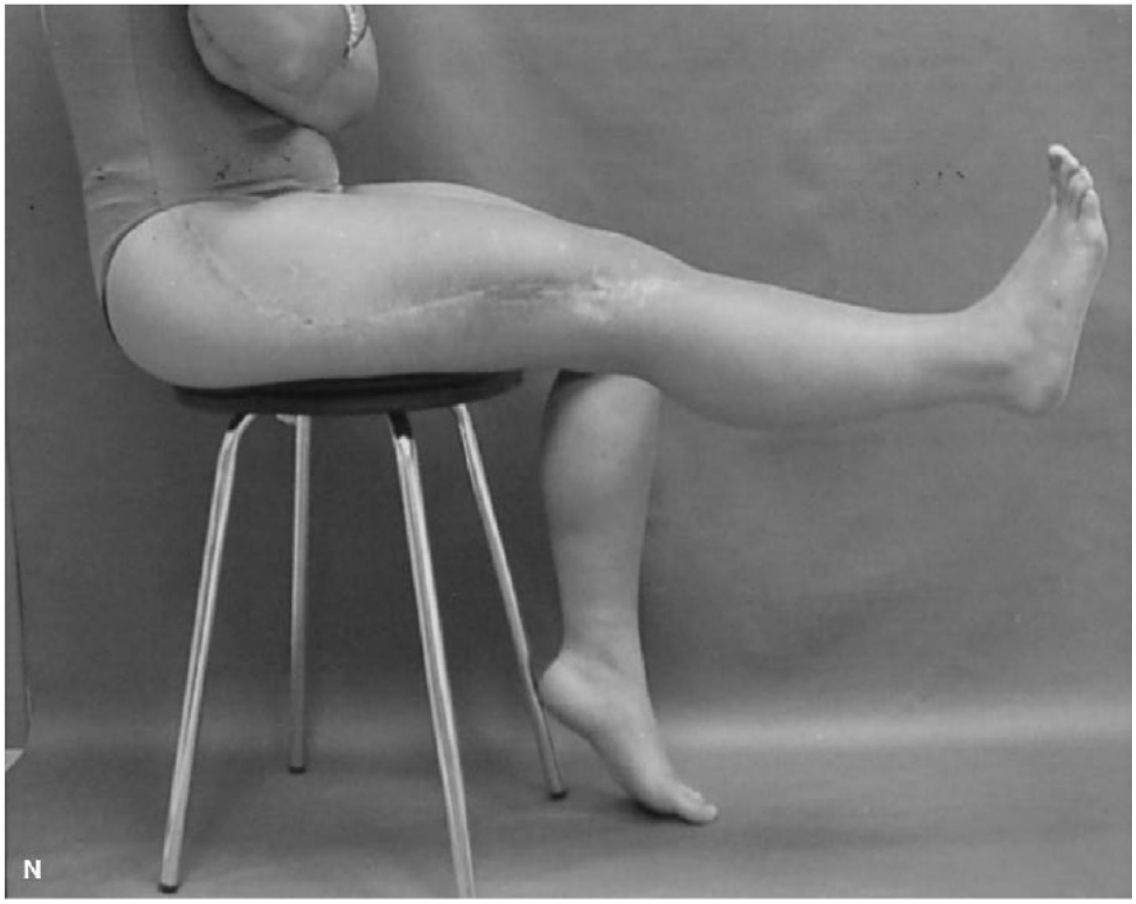
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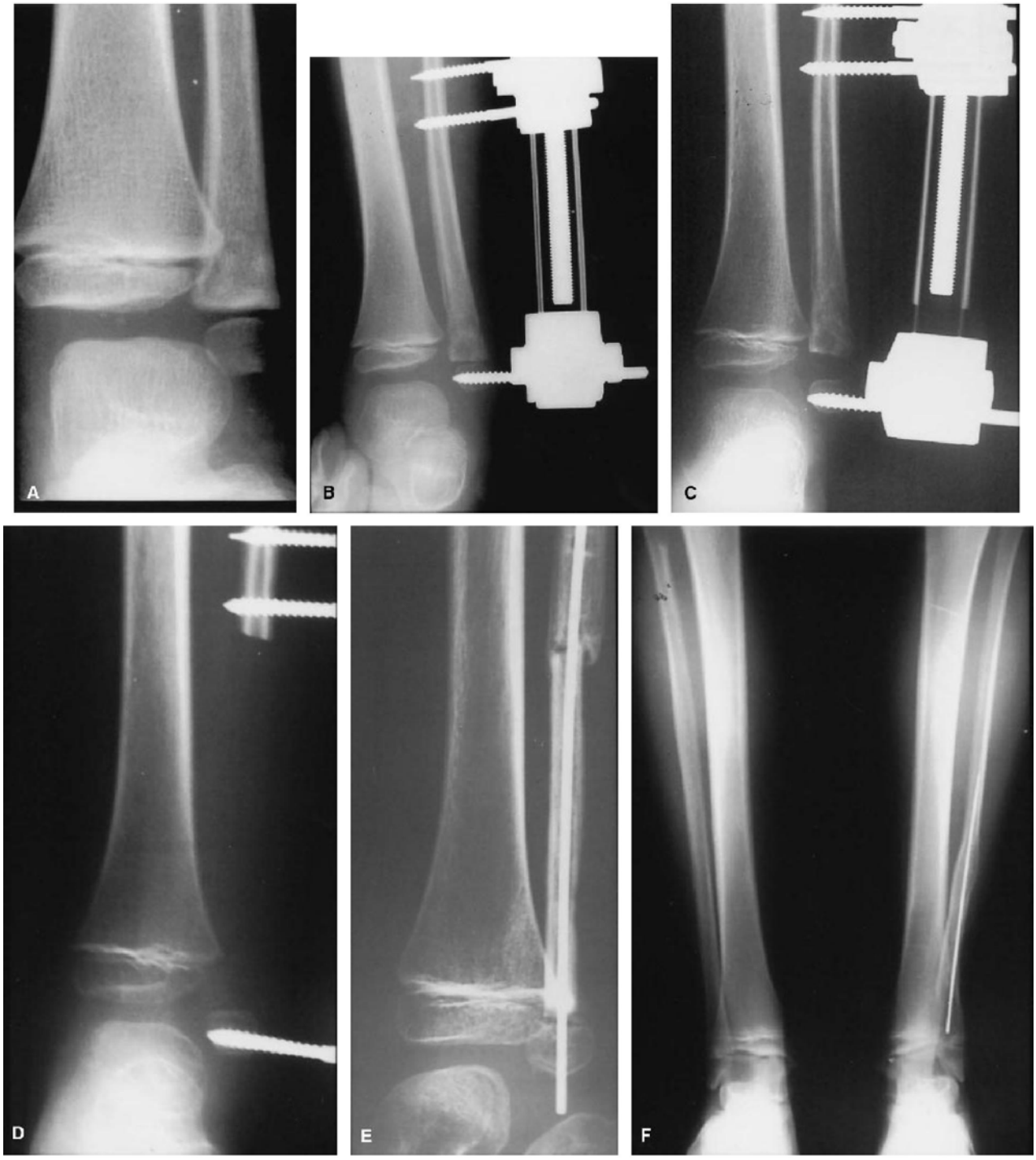
Figure 2. (A) A 4-year-old boy diagnosed with Ewing sarcoma in the distal fibula in 1985. (B–D) Epiphysiolysis before excision of the tumor was performed. (E) Reconstruction was carried out with autologous contralateral nonvascularized fibula. No further operations were required. (F,G) Sixteen years later the patient remains absolutely asymptomatic and plays sports such as soccer. (H, I) Note the excellent function of the ankle joint.

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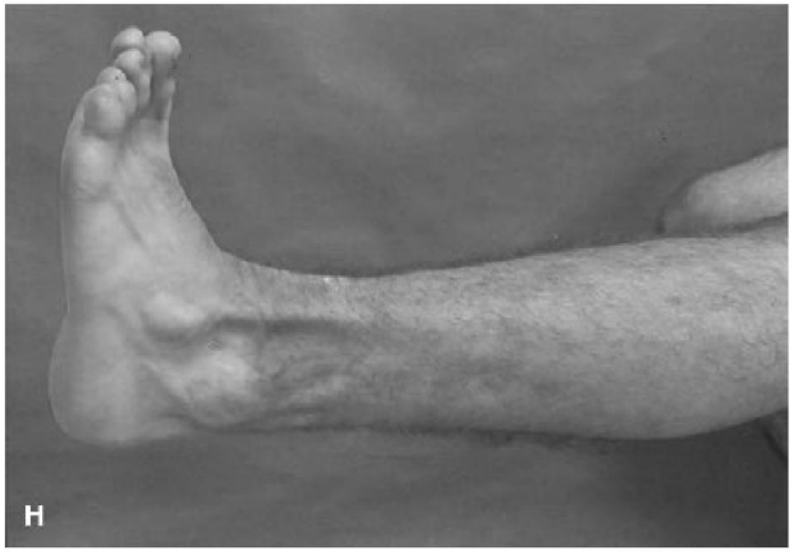
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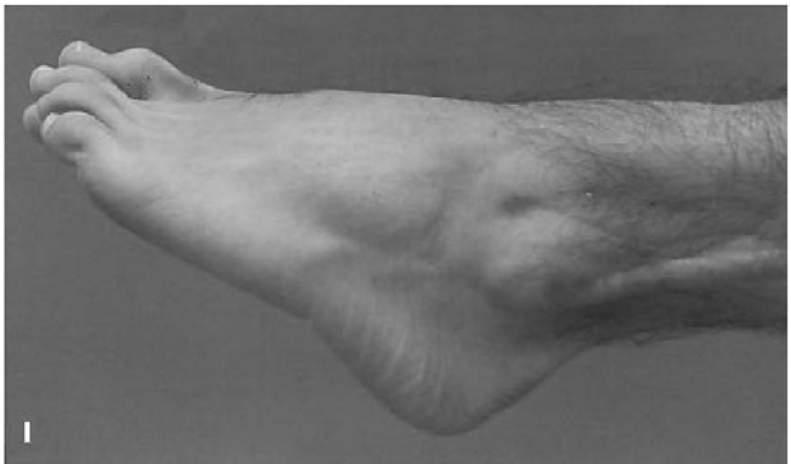
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