Acta Orthop. Belg., 2013, **79**, 239-242

CASE REPORT

Gollop-Wolfgang Complex : an alternative to amputation

Alice NLANDU, Pierre-Louis DOCQUIER

From the Cliniques Universitaires Saint-Luc, Brussels, Belgium

A case of Gollop-Wolfgang syndrome is reported in which an alternative treatment to amputation was chosen. The patient had the classical Y-shaped femur and total tibial agenesis. The recommended treatment in absence of extensor apparatus is knee disarticulation but the parents refused amputation. An alternative treatment was proposed to allow the child to walk. Tibialisation of the fibula and foot positioning under the fibula was performed with good functional result.

Keywords : tibial hemimelia ; Gollop-Wolfgang syndrome ; amputation ; limb salvage.

INTRODUCTION

Gollop-Wolfgang complex is a rare congenital condition which combines a bifurcated distal femur with an agenesis of the ipsilateral tibia. Patients affected with the syndrome may also have an ectrodactyly and present cardiovascular or urogenital anomalies.

The treatment that was proposed for several years to patients with tibia agenesis without a quadriceps was amputation and prosthesis. This solution is very efficient regarding function. Sometimes amputation is not accepted by the parents, for religious or philosophical reasons, or with the hope of medical advances in the future.

We report a case of Gollop-Wolfgang syndrome, managed in our institution without amputation, as the parents were opposed to it. Without any treatment, the syndrome is not compatible with weight bearing on the affected limb.

The patient and her family were informed that data concerning the case would be submitted for publication.

PATIENT AND METHODS

This girl was the first born of a Moroccan family of three children. She was a full-term baby born through cesarean section. Her parents were healthy, without any significant past medical history and were not consanguineous. She had two little brothers. The first one has ectrodactyly of his right hand, whereas the youngest brother does not exhibit any limb anomaly.

This girl had the Gollop-Wolfgang syndrome with the classical limb malformation that was diagnosed antenatally by echography. She had a Y-shaped bifurcated right femur corresponding to a medial and lateral condyle. The ipsilateral tibia was absent, and there was also hypoplasia of the first ray of the foot (Fig. 1).

■ Alice Nlandu, MD, Resident.
■ Pierre-Louis Docquier, MD, PhD, Orthopaedic Surgeon.
Department of Orthopaedic Surgery, Cliniques universitaires St-Luc, Bruxelles, Belgium.
Correspondence : Pierre-Louis Docquier, Department of orthopaedic surgery, Cliniques Universitaires Saint-Luc, 10, avenue Hippocrate, B-1200 Bruxelles, Belgique.
E-mail : Pierre-Louis.Docquier@uclouvain.be

No benefits or funds were received in support of this study.
The authors report no conflict of interests.

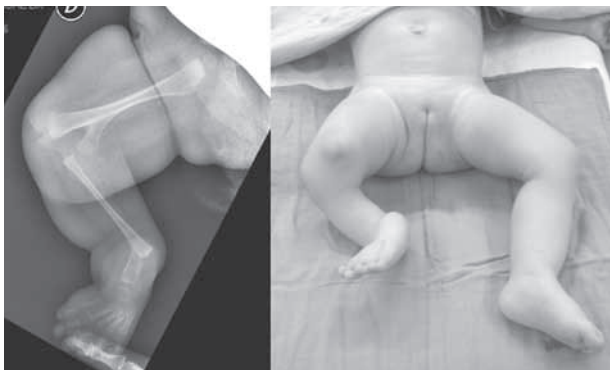

Fig. 1. — Preoperative radiograph and picture of the 8-month-old girl showing the typical Y-shaped bifurcated femur, with tibial agenesis and hypoplasia of the first ray of the foot.

The first treatment proposed was an amputation followed by prosthesis. This option was not accepted by the parents, due to religious reasons. Another option was chosen to allow the child to walk. Before undergoing any surgery, we checked the functionality of the knee. Clinical evaluation and echography did not show any activity of the quadriceps muscle. There was no patella, nor tibia remnant.

We performed surgery in two steps : tibialisation of the fibula at the age of 8 months and foot positioning under the fibula at the age of 12 months. At the time of the first surgical procedure, the bifurcated part of the femur corresponding to the medial condyle was removed. The fibula was positioned under the femur and a chondrodesis was made between the distal femoral epiphysis and the proximal fibular epiphysis with fixation with an intramedullary Kirschner wire (Fig. 2). An above-knee cast was maintained for 6 weeks.

At the time of the second surgery, we performed a chondrodesis of the talus with the distal fibular epiphysis. Fixation was performed with a Kirschner wire (femoro-fibulo-talar K-wire) with the foot in equinus position to facilitate prosthesis fitting (Fig. 2). A six-week cast immobilization was maintained with a walking cast for the two last weeks.

At 13 months of age, she began to walk with a custom-made prosthesis. The prosthesis was changed every 6 months until the age of 2 years, and then yearly to adapt for growth.

At the age of 19 months, she broke the femoro-fibular K-wire, which was removed except for its fibulo-talar part.

At the age of 6 years, she sustained a distal fibular fracture which healed with closed reduction and cast immobilisation. She also broke the fibula-talar wire but this was not symptomatic.

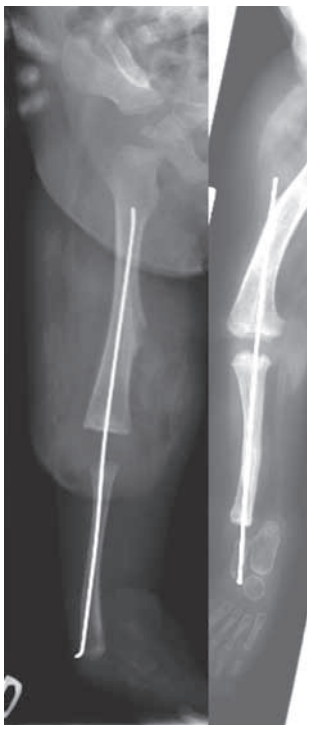

Fig. 2. — Left : first operative step : femoro-fibular chondrodesis with intramedullary nailing. Right : second operative step : positioning of the foot under the fibula with talo-fibular chondrodesis with nailing.

At the age of 9 years and 10 months the wire of the foot was removed because of ankle pain.

The child is now very functional with her prosthesis (Fig. 3 & 4). She is able to walk, to run, to do gymnastics at school, but she cannot bend her knee. The chondrodesis of the knee and ankle did not fuse and there is a slight mobility in the knee and ankle, which is not painful (Fig. 5).

DISCUSSION

The eponym «Gollop-Wolfgang complex» or «bifurcated femora-hand ectrodactyly» was introduced in 1986 by Lurie and Ilyina (*7*). Previously Gollop *et al* described in 1980 two brothers with ectrodactyly and unilateral bifurcation of the femur, absence of both tibiae and monodactyly of the feet. In 1984, Wolfgang reported a case of unilateral

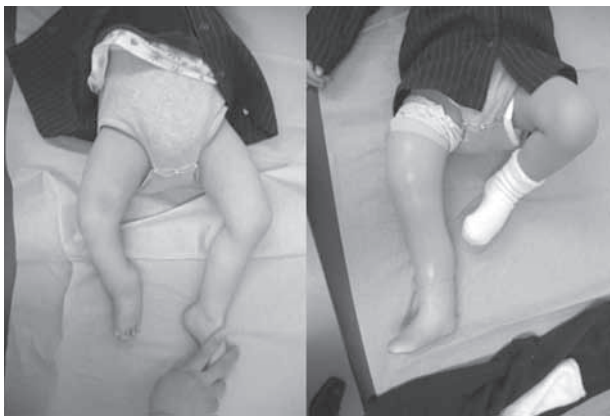

Fig. 3. — Picture of the child and her prosthesis at the age of two years.

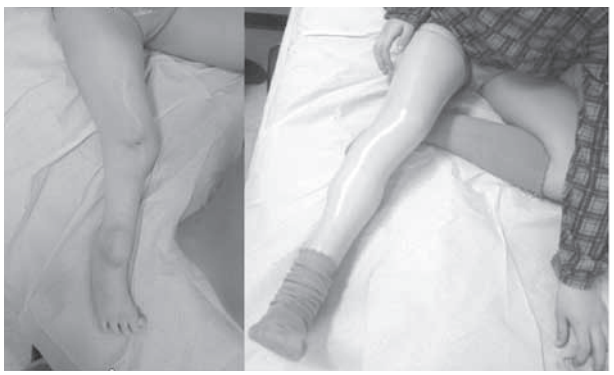

Fig. 4. — Picture of the child and her prosthesis at the age of eight years.

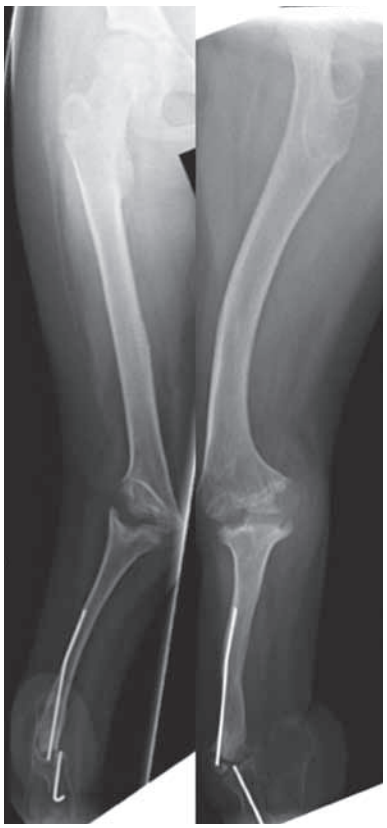

Fig. 5. — Radiograph of the leg at the age of eight years showing no fusion of the femorofibular chondrodesis nor of the fibulotalar chondrodesis.

femoral bifurcation and absence of the tibia (*16*). Few publications about the Gollop-Wolfgang syndrome appeared in the medical genetics literature (*1, 9,14*) and in the orthopaedic literature (*3,6,12,13,15*).

Femoral bifurcation in combination with tibial aplasia but without hand ectrodactyly, as well as tibial agenesis-ectrodactyly syndrome represent a variant of the same developmental field defect (*2,4,10,11*).

The aetiology of the Gollop-Wolfgang complex is most likely an error in the complex genetic control of limb development but the exact cause is still unclear (*3*). In the Muslim population the genetic inheritance seems to be autosomal recessive. This hypothesis is based on the fact that consanguinity is high (*10*). In our case, parents were not consanguineous. Nevertheless, they did not wish to undergo any genetic counselling. Cases of association between prenatal exposure to valproic acid and Gollop-Wolgang syndrome have been reported (*1,8*). In our case, the mother was not epileptic and did not take valproic acid.

In children with the Gollop-Wolfgang complex, it is most important to know if there is any tibial anlage. Echography is a good method to evaluate the tibial anlage. If a cartilaginous tibial anlage is identified and quadriceps is present, any possible reconstruction of the lower leg should be considered with a good active extension (*3*). If there is total absence of the tibia without presence of an extensor mechanism (Jones type Ia) (*5*), knee desarticulation and prosthesis is the best course of action (*3*).

In children having sustained knee disarticulation, walking is possible with an above-knee prosthesis. The gait is characterized by circumduction, resembling the gait of a patient with arthrodesis of the knee (*3*). In our case, the result obtained is comparable to knee disarticulation. The child is able to walk, to run and to do gymnastics at school. This could be a good alternative to knee disarticulation, especially in the absence of parental approval.

REFERENCES

1. **Alessandri JL, Isidor B, David A *et al.*** Tibial developmental field defect in valproic acid embryopathy : Report on three cases. *Am J Med Genet A* 2010 ; 152 : 2805-2809.
2. **Bodurtha J, Coutinho M, Benator R *et al.*** Femoral duplication : a case report. *Am J Med Genet* 1989 ; 33 : 165-169.
3. **Bos CFA, Taminiau AHM.** A 5-year follow-up study after knee disarticulation in two cases of Gollop-Wolfgang complex. *J Pediatr Orthop B* 2007 ; 16 : 409-413.
4. **Evans JA, Chudley AE.** Tibial agenesis, femoral duplication, and caudal midline anomalies. *Am J Med Genet* 1999 ; 85 : 13-19.
5. **Jones D, Barnes J, Lloyd-Roberts GC.** Congenital aplasia and dysplasia of the tibia with intact fibula. Classification and management. *J Bone Joint Surg* 1978 ; 60-B : 31-39.
6. **Kotakemori K, Ito J.** Femoral bifurcation with tibial aplasia. A case report and review of the literatures. *Clin Orthop Relat Res* 1978 ; 135 : 26-28.
7. **Lurie IW, Ilyina HG.** Gollop-Wolfgang complex in a 3-month-old girl. *Am J Med Genet Suppl* 1986 ; 2 : 191-194.
8. **Mercadal Orfila G, Blasco Mascaró I.** Gollop-Wolfgang Complex in a baby born to an epileptic mother treated with valproic acid during pregnancy. *Farm Hosp* 2010 ; 34 : 149-151.
9. **Okumus N, Uner C.** Tibial agenesis and Gollop-Wolfgang complex. *Am J Med Genet A* 2007 ; 143-A : 2490.
10. **Raas-Rothschild A, Nir A, Ergaz Z, Bar Ziv J, Rein AJ.** Agenesis of tibia with ectrodactyly/Gollop-Wolfgang complex associated with congenital heart malformations and additional skeletal abnormalities. *Am J Med Genet* 1999 ; 84 : 361-364.
11. **Richieri-Costa A, Brunoni D, Laredo Filho J, Kasinski S.** Tibial aplasia-ectrodactyly as variant expression of the Gollop-Wolfgang complex : report of a Brazilian family. *Am J Med Genet* 1987 ; 28 : 971-980.
12. **Schoenecker PL, Capelli AM, Millar EA *et al.*** Congenital longitudinal deficiency of the tibia. *J Bone Joint Surg* 1989 ; 71-A : 278-287.
13. **Ugras AA, Sungur I, Akyildiz MF, Ercin E.** Tibial hemimelia and femoral bifurcation. *Orthopedics* 2010 ; 33 : 124-126.
14. **van de Kamp JM, van der Smagt JJ, Bos CFA *et al.*** Bifurcation of the femur with tibial agenesis and additional anomalies. *Am J Med Genet A* 2005 ; 138 : 45-50.
15. **Wehbé MA, Weinstein SL, Ponseti IV.** Tibial agenesis. *J Pediatr Orthop* 1981 ; 1 : 395-399.
16. **Wolfgang GL.** Complex congenital anomalies of the lower extremities : femoral bifurcation, tibial hemimelia, and diastasis of the ankle. Case report and review of the literature. *J Bone Joint Surg* 1984 ; 66-A : 453-458.