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Treatment of Congenital Pseudoarthrosis with Intramedullary Rod in Combination with either Vascularized Fibular Graft or Ilizarov Method

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Abstract : Congenital Pseudoarthrosis of Tibia (CPT) is a rare disease and difficult to treat. The most accepted treatment methods is complete excision of disease tissue followed by either intramedullary nail (IMN) with cancellous graft, vascularized fibular graft (VFG) or ilizarov bone transport.

We review result of treating CPT with combination of IMN with either VFG or Ilizarov method. Visualised fibular graft was used to bridge the gap of more than 4 centimeter (cm) following resection of disease tissue whilst Ilizarov technique was applied when the gap was 4 cm or less. In the VFG group, IMN was inserted after the fibula became hypertrophy to treat distal junctional nonunion. However, in the Ilizarov group, IMN was inserted in the beginning of reconstruction.

There were 3 patients treated with VFG and 2 patients treated with Ilizarov method. All patients had united tibia, with one awaiting intramedullary nailing. There were clinical evidence of neurofibromatosis type I (NF1) in all patients. However, there was no evidence of neural tissue in all excised specimen.

Combination of IMN fixation with VFG or Ilizarov as primary treatment of CPT is a safe alternative treatment method to achieve union and prevent recurrent fracture.

Introduction

CPT is a rare problem and poses a challenge to the treatment. It presents with varieties of severity and has been treated at different age at different centers. A true success of CPT treatment can only be known by following the patients until maturity[2].

It is rather difficult to conclude which method is the treatment of choice. The most accepted treatment method is complete excision of disease tissue followed by either intramedullary nail (IMN) with cancellous bone graft, vascularised fibular graft (VFG) or Ilizarov bone transport. Despite apparently solid clinical and radiographical union recurrent fracture is common[4)7)11)]. We

Key words : congenital pseudoarthrosis, tibia, vascularised fibular graft, ilizarov, intramedullary nail

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Table 1.

case	sex	NF1	type	Previous Tx	Shortening	Age Tx	Present of nerve	Bone gap	Type Reconstruction	Further Tx	result
P1	M	Yes	Ⅳ	No	2 cm	3	nil	5.5 cm	VFG	nail	united
P2	M	Yes	Ⅳ	No	2 cm	5	nil	10 cm	VFG	nail	united
P3	M	Yes	Ⅳ	Yes	6 cm	11	nil	7 cm	VFG	Waiting for nail and lengtheing	united
P4	M	Yes	Ⅳ	Yes	7 cm	8	nil	3.5 cm	Ilizarov + nail	BG	united
P5	f	Yes	Ⅳ	No	3 cm	3	nil	3.5 cm	Ilizarov + nail	no	united

NF1 : presence of Neurofibromatosis type 1. type : base on Crawford Classification. Shortening : leg length discrepancy at presentation. VFG : vascularised fibular graft. Tx : treatment

review our experiences of using Rush rod in combination with either VFG or Ilizarov method in treating CPT at Hospital Universiti Sains Malaysia (USM).

Methods and surgical procedures

We retrospectively reviewed records and radiograph of 5 patients with CPT treated in HUSM since 1988 till 2005. The patient's age, sex, feature of NF1, type of CPT, surgical procedure, tissue histopathological examination, complication, and union were reviewed.

Surgical procedures :

All patients underwent a complete resection of diseased tissue composed of bone, periosteum and fibrous tissue. A bony defect of 4 cm or less was reconstructed using a combination of IMN and Ilizarov bone transport. Reaming was done from osteotomy site down to distal tibia, talus and calcaneum. Rush nail was inserted from the heel, calcaneum, talus, distal tibia passing through the gap to the proximal tibia. Corticotomy was done at the proximal tibia. In case 4 (Table 1), middle segment of tibia was gradually transported down to dock with the distal part. Once docking was achieved, distraction was continued from proximal to corticotomy site to lengthen the leg. In case 5 (Table 1), the leg was shorten to produce a primary docking. The distraction was done from proximal tibia corticotomy to lengthen the leg. In both cases, the ilizarov ring fixators were removed after the desired length was achieved, and the legs were protected with long leg cast until consolidation.

Patients with the skeletal defect of more than 4 cm were reconstructed using vascularised fibula graft. The graft was stabilized by inserting its both ends to medullary cavity of recipient bone and transfixed with wires or plate and screws. Externally, the leg were supported with long leg cast until union. In the first two cases we observed junctional nonunion and recurrent fracture. This problem was treated with intramedullary Rush rod at 6 months when the fibular graft hypertrophy.

Results

There were 5 patients, ages between 3 to 11 years old. One of them was female and 4 were males. All were classified as Crawford type Ⅳ[3]. Clinical evidence of NF I were present in all patients. The resected diseased tissues were examined histologically. There was no nerve tissue found in resected specimen from all patients. The defective gaps were between 3 cm to 10 cm.

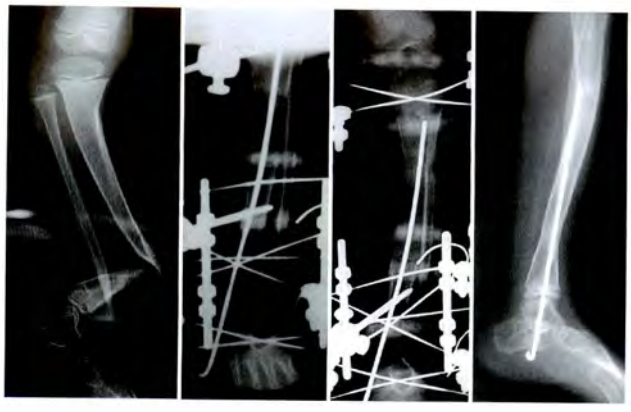
Three patients (P1, P2, P3) were treated with VFG. The first two patients had proximal junctional union but developed nonunion of distal junctional zone (Fig. 1). Union of the distal junction was achieved after intramedullary rush rod insertion. Early results of these two cases

a | b | c **Fig. 1.**

a : Radiograph of patient 2 before undergoing resection anf VFG reconstruction

b : Vascularised fibular graft that united proximally with nonunion of distal junctional zone treated with intramedullary nail

c : Solid union with intramedullary nail in situ



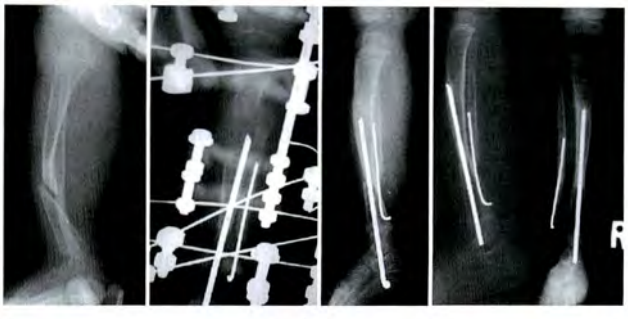
a | b | c | d **Fig. 2.** Serial Radiograph of Patient 4

a : Ragdiograph of P4 before undergoing resection and bone transport

b : Radiograph showing transported middle segment before docking

c : Union of docking area was assisted by cancellous bone graft

d : A solid union with rush rod insitu



a | b | c | d **Fig. 3.** Serial Radiograph of patient 5

a : Radiograph at age of 18 months old befsection, primary docking and limb lengthening

b : Radiograph at the end of distraction

c : Union achieved at 3 months

d : 8months after removal of tip of rush rod showing a solid union with growth of distal tibia

were already published[17]. The third patient has evidence of union. The IMN will be inserted once the fibula become hypertrophy.

Two patients (P4 and P5) had resection, intamedullary rod, distraction histiogenesis. Patient P4, who underwent delayed docking with bone transport followed by limb lengthening, needed secondary bone graft procedure to assist union at docking site (Fig. 2). Patient P5 who underwent primary docking and distraction histiogenesis for limb lengthening (Fig. 3) had union without any further procedure.

Discussion

CPT was found to be associated with NF1 in 40%[16] to 77% of cases[9]. However, signs of NFI were present in all our series. The pathologic process of CPT is the growth of abnormal, fibromatosis-like tissue either within the periosteum (dysplastic type) or within the endosteal/marrow tissue (cystic type) or coexistence of both (mixed type)[6][9]. Examination of excised tissue from diseased site did not show any evidence of neural tissue and thus is consistent with other reports[9].

A complete excision of the diseased tissue is necessary in treating CPT and it is evidenced by visualizing normal bleeding and marrow cavity[6]. Resection leaves bone gaps between 1 to 3 cm[4]. The bone gaps in our series varied from 3 to 10 cm. Resection of diseased tissue followed by fragment approximation, intramedullary rodding and bone graft is relatively easy procedure, thus,

Joseph B et al suggests rodding as initial treatment of CPT[10]. The union rate achieved with this technique was 86%[10]. Simonis et al suggests VFG as a primary treatment when the gap was more than 3 cm[15]. We tailored our treatment based on the bony gap created. Preoperatively, level of medullary cavity on plain radiograph can be used to estimate the reconstructive method. Eventually, intra-operative measurement of the bony gap would determine the type of definitive reconstruction[6].

Reconstruction by primary docking with Ilizarov method (case P5) produced union without the need of further procedure. It has the advantage of healing like a fresh fracture. However, fibrous tissue interposition may disturb the union process in delayed docking procedure. Thus, case P4 required further bone grafting to facilitate union. The presence of intramedullary device has a role of guiding bone transport and docking. It also maintains alignment and provides load sharing protection for axial force allowing micromotion to enhance union. Intramedullary nail crossing the corticotomy site help to decrease time for wearing external fixator while waiting for consolidation[13]. Reaming of endosteal blood supply did not prolong time taken for consolidation of newly formed bone[13].

Reconstruction by using vascularised fibular graft can replace a large bony defect. When VFG was used, it had a reported success rates between 92%-100%[5][8]. In first two cases, we observed that distal functional nonunion. We think it could be due to unstable fixation near to the ankle joint. Dormans et al reported the use of intramedullary rod and bone grafting to treat recurrent fractures associated with vascularised fibular graft[5]. The intramedullary rod was recommended after fibula become hypertrophy[5]. In our cases, union was achieved after insertion of intramedulary rod without requiring further additional bone graft. The success of this treatment was attributed to a good stability and alignment provided by intramedullary rod. Furthermore, adequate resection of the diseased bone and periosteum had been replaced by a healthy vascularised fibula graft. We propose the insertion of intramedullary nail in all cases reconstructed with VFG upon its hypertrophy.

We used rush rods in all cases and kept the ankles transfixed. It produces some disability from ankle stiffness. However, the stiffness produces a very stable construct especially for a very distal lesion and prevents valgus deformity of the ankle. Dobbs et al recommend the use of Wiliam Rod to minimize ankle stiffness[4]. Longitudinal growth of tibia pushes distal part of tibia and foot to migrate distally but a straight William rod will stay in place leaving the ankle free[4]. However, in the presence of concomitant fibula pseudoathrosis, valgus ankle deformity may appear upon freeing of ankle following longitudinal growth of tibia. Thus, it was suggested for placement of syndesmotic screw to solve the problem of valgus ankle[4]. Intramedullary rod traversing the growth plate of distal tibia is less likely to cause deformity of distal tibia if it is located at the centre[4].

Treatment can be as early as 2 years with intramedullary nail, vascularized fibula graft or Ilizarov methods[1][10][14]. Boero et al reported that patients with age more than 5 years old consolidate better with Ilizarov method[1][2]. We prefer to initiate treatment early in life to avoid the sequalae of long standing problem like severe shortening and disuse osteoporosis (Table 1). We observe that, leg length discrepancy was more in older patient (Table 1). This is consistent with

reported series by Boero et al[1]. Leg length discrepancy in CPT is a result of chronic resorption of bone at pseudoarhtrosis site prior to treatment[4]. Morrissy suggeated that shortening was due to inhibition of growth of distal tibial physis[12].

Conclusion

Our experience showed that complete resection of disease tissue facilitate union. Reconstructions with Ilizarov bone transport for bony gap of 4 cm or less and vascularized fibula graft for those more than 4 cm were safe treatment options. Intramedullary nail is necessary to enhance union and prevent recurrent fracture in both procedures.

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