Original Article

Patterns of Presentation and Outcome of Management of Congenital Pesudarthrosis of the Tibia

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ABSTRACT

Background: Congenital pseudarthrosis of the tibia (CPT) is a rare paediatric orthopaedics condition characterized by bowing of the tibia with spontaneous fracture which does not heal easily. It is sometimes associated with Neurofibromatosis. It has many classification schemes and various methods of treatment.

Objectives: To study the patterns of presentation, demographic characteristic and outcome of management of CPT.

Patients and Methods: This is retrospective case series study, records and follow up notes of all patients of CPT who presented to and managed at Soba University Hospital between 2003 and 2014 were reviewed, data were collected and analysed.

Results: there were 37 patients with CPT, 19 males and 18 females. Their ages at presentation ranged between 1month and 11 year, mean of 3.8 years. Neurofibromatosis was associated in 37.8% of the cases. The most common deformity was anterolateral bowing of the tibia seen in over 90% of the cases. In 54% of cases the treatment was surgical and 90% of these had intramedullary rush pin and free fibular grafting. In 88% complete healing could be attained. In two cases there was non-union.

Conclusion: Congenital Pseudarthrosis of the tibia is a rare pathology we have reported 37cases. One third of the patients had Neurofibromatosis. Majority of cases had anterolateral bowing. Associated fibular pseudarthrosis is very rare. If they present before fracture they can be protected by bracing. Intramedullary rush pin with bone grafting is easy, cost effective with union achieved in over 80% of cases.

Key words: Bowing of the tibia, spontaneous fracture**,** neurofibromatosis, Sudan.

Congenital Pesudarthrosis of the tibia (CPT), is rare orthopaedic condition with reported incidence of 1:140,000 to 1:250,000 neonate[1]. It was first described by Paget in 1891 and is characterized by a spontaneous fracture that poorly heals with routine treatment[2].It is usually unilateral and there is no sex predominance and is characterized by anterolateral bowing of the tibia that can progress to pseudarthrosis[3]. Posteromedial bowing of the tibia is benign variant of the tibial deformities which improve with time and does not progress to CPT like the anterolateral bowing. A skin dimpling is seen over the tibia[4]. Anteromedial bowing of the tibia is reported to be associated with fibular hemimelia [2] and fibrous dysplasia[5].

The association of CPT with neurofibromatosis (NF-1) is known and was first reported by Ducroquet in 1937 [2]. Ten percent of patients with neurofibromatosis patients will develop CPT, in 50% of cases CPT is associated neurofibromatosis[3]. There is little evidence for the role of hereditary factor in CPT but there are reports about CPT in familial pattern[6].

An associated pseudarthrosis of the fibula is also seen in most patients with CPT[2].

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The exact cause of the CPT and its site remains unexplained. However poor vascularity, soft tissue interposition and lack of osteoblastic activity were incriminated[7-9].

Pathological findings in CPT include; a thick cuff of poorly perfused hamartomatous fibrous tissue surrounding the Pesudarthrosis site[9]. Clinical presentations range from simple anterolateral bowing to frank Pesudarthrosis[3]. Simple radiological examination can show variety of features; ranging from simple bowing with no medullary cavity, cystic lesion, complete or incomplete fracture of the tibia with atrophic tapered end. But MRI can provide better detailed picture [1].

Age of the patient and whether or not there is a fracture at the time of presentation will affect the treatment. Before walking, there is no treatment but once the child starts to walk orthotic protection is indicated till skeletal maturity[7, 10].

The main aim of management of CPT is to obtain union, prevent re-fracture, equalize limb length, and prevent and correct deformities of leg and ankle and this is achieved by excising the pathological area of the Pesudarthrosis, solid mechanical fixation by intramedullary rod and bone grafting[3, 11-13]. Reconstruction of the area using free vascularized fibular graft was also performed by some authors[12], while others used Ilizarov fixation[7]. Recently bone morphogenetic protein type 2(BMGP2) was used as adjunct to fixation with good results[14].

These new techniques need to be confirmed and the final prognosis remains poor and no treatment can guarantee healing and in fact no method proved superior[3, 9]. Poor prognostic features include associated neurofibromatosis, associated fibular pseudarthrosis [2].

Complications of surgical treatment of CPT include stiffness of ankle, hind-foot valgus, ankle deformity, re-fracture and tibial shortening[3].

Recently many modalities of treatment were introduced, they are very expensive and some are demanding. These modalities may not be available in underprivileged areas where health services are substandard. Thus using rush pin and fibular graft may still be a valid option.

The main objective of this work is to study the demographic characteristics, patterns of presentations and outcome of management of CPT treated at the paediatric orthopaedics department at Soba University Hospital.

PATIENTS AND METHODS:

In this retrospective study, all records of patients of CPT who were treated at Soba University Hospital were reviewed. There were37 patients of CPT, presented and managed between 2003 and 2014. All patients were included and the records including x-rays, operations and follow up notes were reviewed. The procedure of the intramedullary rush pin when performed, was introduced antegrade and non-vascularized fibular graft was used[7]. All patients were managed or operated on by the same surgeon (first author). Demographic characteristics, clinical presentation and management were studied and data collected were analysed using (Statistical Package for Social Sciences (SPSS).

The study was approved by the ethical and research committee of the hospital and hospital authorities were informed and their approval was attained.

RESULTS:

There were 37 patients with CPT, 19 (51.4%) were males. Their ages ranged between one month and 11 years, mean of 3.8 years (Table 1). Twenty five patients (67.6%) presented below the age of four years. Sixteen patients (84.2%) were more

than three years of age when operated on and three patients (15.8%) were below three years of age. Fourteen patients (37.8%) had associated neurofibromatosis which was diagnosed on clinical basis. Of the 37 patients, 35(94.6%) had anterolateral bowing. 25 patients (67.6%) had established pseudarthrosis (Table 2 and 3).

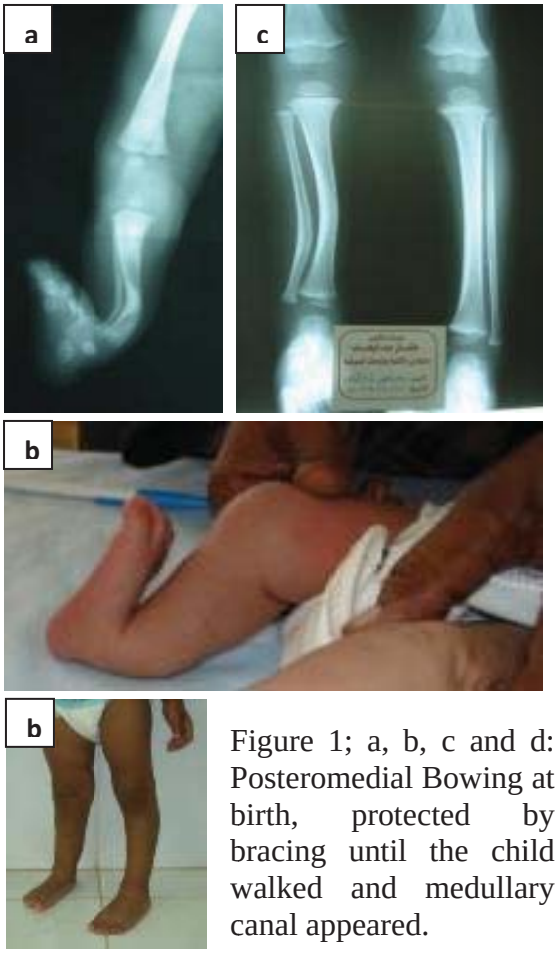

Figure 1; a, b, c and d: Posteromedial Bowing at birth, protected by bracing until the child walked and medullary canal appeared.

Conventional X-ray was the only imaging investigation performed.

Twenty patients (54.05%) were treated surgically; 18 with excision, intramedullary nailing and bone grafting. One of the operated on patients was a revision operation (the primary surgery was done in another hospital) Figure 3 a, b, c and d. One patient had Ilizarov procedure and one underwent amputation (Table 4). Complete healing was achieved in 16 patients (88.88%) of the 18 patients treated with IMN and bone grafting, and the only one patient treated with Ilizarov. Two patients had non-union. Postoperative complications included shortening in seven patients (42.1%) and infection in one patient (Table 4).

Seventeen patients were managed with bracing; 12 of whom had no fracture at presentation and were protected (Figure 1; a, b, c, and d).

Figure 2; a, b, c, d and e: Anterolateral Bowing at birth, protected by bracing.

DISCUSSION:

Congenital pseudarthrosis of the tibia (CPT) is one of most difficult paediatric orthopaedic conditions to treat[3, 14]. It was first described in 1891[2] but association with type 1 neurofibromatosis was first mentioned in 1937[1]. In this study we had

Table 1: Age group distribution of the studied patients (n=37)

Age group in Months	Number of Patients and (%)
0 – 4	25 (67.6)
5- 8	9 (24.3)
9 – 12	3 (8.1)
Total	37 (100)

Table 2: Gender, age at presentation and age at surgery in the studied patients with congenital Pseudarthrosis of the tibia.

Demographic characteristics	Number of patients & (%)
Gender (n=37)	
Males	19 (51.4%)
Females	18 (48.6%)
Age at Presentation (n=37)	
< four years	12 (32.4%)
> four years	25 (67.6%)
Age at the time of surgery (n= 19)	
Older than 3 years	16 (84.2%)
Less than 3 years	3 (15.8%)

37 patients with CPT, there was no sex predominance; 18 males and 19 females, a finding that is supported by many other authors [3, 5].

Reports described clinical presentation to range from simple anterolateral bowing of tibia to frank pseudarthrosis[3].In this series 25 out of 37(60%) presented with established pseudarthrosis, while 10 out of 37(27%) presented with bowing. In nearly 95% of cases (35out of37), the deformity was anterolateral. A similar result was reported by Mahnken[1].

In this study Neurofibromatosis (NF-1) was found to be associated with CPT in 14 patients out of 37 (38.8%). Similar findings were reported by other series [(8,11)]. In fact, the association of neurofibromatosis ranged in different series from 40 – 80% of CPT cases [(1)]. The association was higher in the series reported by McKeown and Morrissy (50%)[15,16]. Morrissy stated that 50% of patients with CPT have neurofibromatosis while 5-10% of patients with neurofibromatosis have CPT[16].

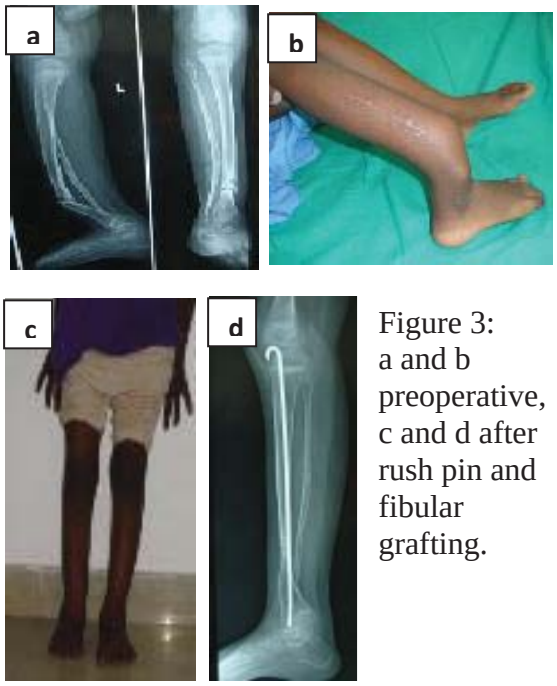

Figure 3: a and b preoperative, c and d after rush pin and fibular grafting.

Table 3: Patterns of presentation and type of deformity in the studied patients (n=37)

Pattern of Presentation	Number of patients & (%)
Established Pseudarthrosis	25 (67.6%)
Bowing	10 (27%)
Shortening	2 (5.4%)
Associated Neurofibromatosis	14 (37. %)
Anterolateral bowing	35(94.6%)
Antero-medial bowing	1 (2.7%)
Postero-medial bowing	1 (2.7%)
Associated Fibular Pseudarthrosis	2 (5.4%)

In this study 17 patients (45.9%) were treated with bracing in 12 of these, bracing, was indicated because they presented with bowing before the fractures. Posteromedial bowing of the tibia is a benign condition that rarely progresses to fracture[7]. In this series posteromedial bowing responded well to

bracing (Fig 1 a, b, c and d), and another case of anterolateral bowing was managed by bracing (Figure 2 a, b and c). Managing patients with bracing before fracture occurs was advised by many authors [7, 10]. Sixteen out of the 18 patients (88.88%) treated using intramedullary rush pin and bone grafting had good healing, this is similar to reports by Shah[12] and Bakers[17]. The latter in his series attained union in 87% of patients using intramedullary nail fixation bone grafting and electric stimulation, and 60% when only intramedullary fixation and bone-grafting was used. In the literature the rate of union varies from 28% to 80% [3].

Table 4: Type of Surgery and results in surgically treated patients (n=20)

Type of Surgery	No of Patients	Results			
		Union	Non union	Infection	Shortening
IMN and bone grafting	18	16 (88.89%)	2 (11.11%)	1 (5.56%)	6 (33.33%)
Ilizarov fixation	1	1	--		1
Amputation	1	--	--	--	--
Total	20	17	2	1	7

CONCLUSION:

Congenital Pseudarthrosis of the Tibia is a rare pathology. One third of patients will have an associated Neurofibromatosis. It presents in majority of cases as anterolateral bowing and very rarely there is concomitant fibular pseudarthrosis. If they present before fracture they can be protected by bracing. Intramedullary rush pin with bone grafting is easy, cost effective with union achieved in over 80% of cases.

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