Ponseti Method in the Management of Syndromic and Arthrogrypotic Clubfoot: Outcome of Treatment in Iraqi Patients

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ABSTRACT:

BACKGROUND:

Clubfoot is one of the most common congenital birth defects, most occuras an isolated birth defect and are considered idiopathic. The Ponseti method of serial casting has gained widespread popularity in recent years for the treatment of idiopathic clubfoot**.**Non-idiopathic clubfeet are thought to be too rigid to correct by casting alone, it is often treated primarily with surgical extensiveSoft tissuereleases, and the use of Ponseti method for the treatment of non-idiopathic clubfoot has not been extensively reported.

AIM OF THE STUDY:

To study the effectiveness of Ponseti method to achieve correction in non-idiopathic clubfoot, and to follow the patient for relapses.

PATIENTS AND METHOD:

During the period from February 2013 to December 2017, 16 infants with clubfeet associated with other congenital deformities were managed by Ponseti method, and followed for a minimum of 10 months and maximum of 60 months (mean ± SD= 32.94±17.39). All patients were treated at medical city center in Baghdad/Iraq.

RESULTS:

16 patients with syndromic clubfoot, 9 of them diagnosed as arthrogryposis and the rest as other syndromic types. 12 of them were males(75.0%) and 4 of them were females (25.0%). 14 of cases had bilateral deformities (87.5%) and 2 unilateral deformities (12.5%).5 patients required 4- 5 casts to achieve initial correction (31.2%) and 11 patients required 6-8 (68.8%).In arthrogryposis only 55.6% of cases required 5 and more casting, while in other syndromic cases 85.7% of cases required 5 and more casting**.** All patients required additional minor surgical procedures. Initial Pirani score of pre casting to the right and left was (**mean±SD**) 4.83±0.81 and 5.03±0.85 respectively, Pirani score after removal of final cast(pre surgery) to the right and left was (**mean±SD**) 1.03±0.74 and 1.3±0.7 respectively (p –value < 0.001).Follow-up for minimum 10 months and maximum 60 months (**mean±SD**/ 32.94±17.39) .Half of our patients got plantigrade, pain free, and flexible foot at last follow up with no relapse, another half of patients developed relapse at subsequent follow up. Most relapses occurred in arthrogryposis cases (66.7%),while in other syndromic cases relapses occurred in (28.6%).Two of relapsing cases(25%) treated by repeated casting, another two cases(25%) treated by repeated casting and heel cord release, and four cases (50%) treated by salvage procedure (talectomy) after failed repeated casting due to rigid and stiff feet.

CONCLUSION:

The Ponseti method is effective treatment for syndromic clubfootto achieve flexible, pain free and plantigrade foot and to minimize the extensive future surgeries.

KEY WORDS: Syndromic Clubfoot, Ponseti method, Outcome of treatment

INTRODUCTION:

The term '*talipes*' is derived from *talus* (Latin = anklebone) and *pes* (Latin = foot),Equinovarus is the most important talipes deformity [(1)].

Clubfoot occurs in approximately one in 1000 live birthsand is one of the most common congenital birth defects[(2)],It is easily recognizable at birth and can be differentiatedfrom the more common positional footanomalies on the basis of the rigid ankle equinus deformity and resistance to simple passive correction, Most clubfeet occuras an isolated birth defect and are considered idiopathic[(3,4,5)].

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The Ponseti method of serial casting gradually corrects thedeformity, combined with a percutaneous tenotomy of the Achilles tendon for the ankle equinus followed by bracing to maintain the correction, has gained widespread popularity in recent years for the treatment of idiopathic clubfoot[2-5]. When treatment with the Ponseti method is initiated in the first few years of life, patients with idiopathicclubfoot rarely require extensive surgical treatmentand commonly have excellent long-term outcomes[6]

Non-idiopathic clubfoot occurs in patients with additional malformations, chromosomal abnormalities, or known genetic syndromes, such as arthrogryposis, although there have been multiple reports of successful treatment of idiopathic clubfoot with use of the Ponseti method[2-5],on the contrary, non-idiopathic clubfoot is often treated primarily with extensive surgical Soft tissue release as the deformityis thought to be too rigid to correct with casting alone[7].

Arthrogryposis includes a heterogeneous group of disorders characterized by multiple joint contractures, including flexed or extended knees, hip dislocations and upper extremity deformities and clubfeet which tend to be severe, rigid, difficult to correct and has a high recurrence rate [8,9,10]. Managing arthrogrypotic clubfeet has traditionally high failure rate as well as complications[11].

The Ponseti method of manipulation and casting can be useful in rigid, teratogenic clubfeet [2]. Few reports have been published on the results of the Ponseti method in the treatment of syndromic clubfoot[12-14]. In this study, we present our experience in treating syndromic club feet using the Ponsetimethod

PATIENTS AND METHODS:

During the period from February 2013 to December 2017, 16 consecutive infants with clubfeet deformities (14 bilateral clubfeet deformities and 2unilateral) associated with other congenital deformities were managed by Ponseti method, and followed for a minimum of 10 months and maximum of 60 months (mean±SD= 32.94±17.39). All patient were treated at medical city center.

Study protocol was reviewed and granted permission by the Ethical Committee of ministry of health / medical city health directorate. After explaining the objective of the study to the parents, verbal consent was obtained from them prior to intervention. Names of participants were replaced by identification codes to keep data confidential. To carry out the study, official agreement was obtained from medical city health directorate.

All patient underwent assessment by a clinical geneticists and clinical neurologists to confirm the diagnosis of syndromic types. At presentation, demographic data were collected (table 1).All patients parents had been given full explanation about nature of our management. 9 patients were diagnosed as arthrogryposis, all presented with an appearance of classicarthrogryposis (four-extremity involvement including the larger joints) and another 7patients were diagnosed as other syndromic types.

Inclusion criteria: All infants with clubfoot deformity associated with other congenital deformity(syndromic clubfoot).

Exclusion criteria: Idiopathic, postsurgical and Neurogenic clubfeet.

Regarding gender, males stood out in the high rate for foot deformity in comparison to females in a ratio of 3:1.

Table 1: Distribution of the patients by socio-demographic factors (n=16).

Variables	Number	Percentage (%)
Age		
Less than 1 month	7	43.7%
1-6 months	3	18.8%
7-12 months	4	25.0%
More than 12 months	2	12.5%
Total	16	100.0%
Gender		
Male	12	75.0%
Female	4	25.0%
Total	16	100.0%

At the follow-up visits, each clubfoot was assessedbythe Pirani severity scoring system, and the standard Ponseti p rotocol was used with manipulation and casting of the foot (figure 1and 2).At the end of serial casting we performed minor surgeries in form of either elongation of tendon Achilles ,with or without posterior capsule release or tibialis anterior transfer then last castapplied for two to three weeks in (10-20) degree dorsiflexion and full abduction.

Fig. 1: Patient no. 11 and 6 at initial presentation.

Fig. 2: Patient no. 11 during casting.

After successful initial correction , children were placed in Denis Browne splint (figure 3). Brace was worn full-time for three months, followed by wearing it at night and during nap time until 4 years of age (approximately for 14–16 h every day).

Fig. 3: Denis Browne splint.

Children were followed up initially with 3 monthly clinical review for the first 2 years and then 6-monthly reviews.

Data entry and analysis was done using SPSS version 20 computer software (statistical package for social sciences),categorical variables were presented as frequencies and percentages, continuous variables were presented as (mean ±standard deviation). paired t-test was also used to compare the mean between two paired reading . P value of ≤ 0.05 was considered as statistically significant.

RESULT:

Out of 16 patients with syndromic clubfoot, 56.2% had arthrogryposis, while the other cases are syndromic.5 patients required 4- 5 casts to achieve initial correction (31.2%) and 11 patients required 6-8 (68.8%). Two third required heel cord releasealone ,which represented 62.4% of the patients,while 25.0% needed heel cord release plus posterior capsulotomy, those who needed heel cord release plus lateral column shortening andheel cord release plus Tibialis anterior tendon transfer were evenly distributed as 6.3%. Talectomy accounted for half of the relapsed cases (50.0%), while the remaining 50.0% were treated by repeated casting and heel cord release plus casting.The mean Pirani score at the initial visit for right and left foot is 4.83±0.81 and 5.03±0.85 respectively. Regarding Pirani score pre-operative the mean for the right and left foot is 1.03±0.74 , 1.3±0.7 respectively. For relapse cases Pirani score for the right and left foot is 2.2 ±0.97 and 1.93±1.42 respectively

Figure 4 shows that the distribution of the patients with syndromic clubfoot by prognosis was identical in terms of whether relapse occurred or not.

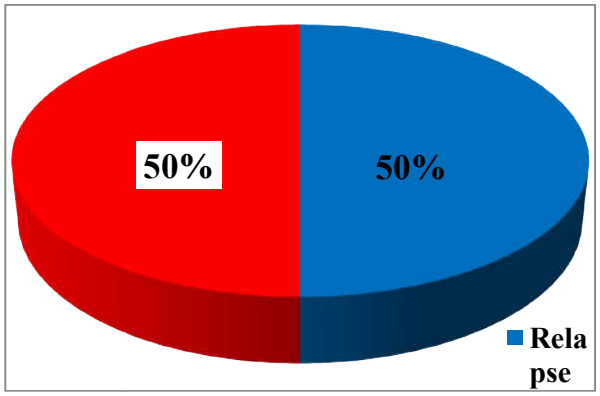

Figure 4: Distribution of the patients by prognosis (n=16).

Table 2 shows that paired t- test was conducted to show the mean differences of Pirani score at initial visit and pre-operative in both right and left deformed foot, There is a significant mean difference in both circumstances (p-value<0.001)

Table 2: Mean Differences of the Pirani Score (initially and pre-operative) of the patients with syndromic clubfoot (n=15).

Study Variable	Study group	N	Mean ± SD	Paired t-test	P value
Pirani Score	Initially right	15	4.83±0.81		
	Pre-operative right	15	1.03±0.74	19	<0.001*
	Initially left	15	5.03±0.85	17.61	<0.001*
	Pre-operative left	15	1.3±0.7		

*P value ≤ 0.05 was significant

The prognosis in arthrogryposis type is relapse in 66.7% while no relapse in 33.3%,regarding other syndromic cases, relapse represented 28.6% while no relapse occur in 71.4%, the result of relapsed cases of arthrogryposis type is talectomy (66.6%),while in other syndromic cases is 50.0% repeated cast and the other 50.0% is repeating heel cord release plus cast, 44.4% of arthrogryposis type of syndromic clubfoot needed less than five casts, while those who

needed six and more casts represented 55.6%, Regarding other syndromic cases, patients needed 4-5 casts in a percentage of 14.3%,while those who needed 6-8 represented 85.7% .

Figure 5 show the Percent satisfaction of our study at final follow up, which include patient not end by salvage procedure(talectomy).

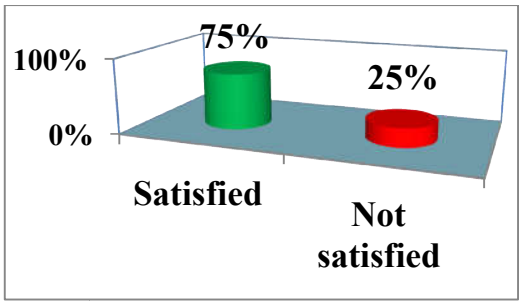

Figure 5: Show percent satisfaction at final follow up.

DISCUSSION:

Treatment of clubfoot is challenging, extensive research has been made regarding the etiology(15-20), surgical treatment(21,22), relapses(23,24), andcasting methods[25].

Excellent results had been documented with the use of Ponseti method of serial casting for patients with idiopathic clubfoot, and reduce the need for more extensive surgery this concept can be applied to syndromic clubfeet[26]. Syndromic Clubfoot tends to be severe, rigid, difficult to correct and has a high recurrence rate**[12].**

Morcuende et al.who treated 16 patients with bilateral syndromic clubfeet, needed as average 7 number of casting to achieve initial correction[14].Boehm et al who treated 12 patients with 24 syndromic clubfeet, required average of6.9 ± 2.1 casts to achieve correction[12].Kowalczyk and Lejman also reported on the short-term results in five patients with 10 arthrogrypotic clubfeet treated by Ponseti method required average of 8.4 casts to get correction[13]. Van Bosse et al. who treated 10 patients with 19 arthrogrypotic clubfeet, needed average of 7.7 cast (range 4-12) to get initial correction by modified Ponseti method which start by heel cord release before casting to unlock calcaneus from posterior tibia and followed by serial casting[27].Hosam E. Matar et al who treated 10 patients with 17 syndromic clubfeet, needed average of 8 casts to get correction (range 4-10)(28/59).this study shows that 5 cases (31.2%) of our patients needed 4- 5 casts and 11cases(68.8%) needed 6-8 casts.

Morcuende et al. only 15 of 16 patients required only heel cord release at the end of casting and one patient required heel cord release plus posterior medial capsule release (PMR)[14]. Boehm et al. requiredheel cord release to all patients to get equinus correction[12].Kowalczyk and Lejmanrequired heel cord release to correct equinus in all patients[13]. Van bosse et al. usedmodified Ponseti method, 10 of 19 feet required second heel cord release at the end of casting(58).Hosam E. Matar et al performed heel cord release in 16 of 17 feet[28].In our patients 62.4% requiredheel cord release alone, while 25.0% needed heel cord release plus posterior capsulotomy, those who needed heel cord release plus lateral column shortening andheel cord release plus Tibialis anterior tendon transfer were evenly distributed as 6.3%.

Morcuende et al had only 4 of 16 patient with relapse,subsequent surgery for relapses (1 bilateral PMR with a repeat left PMR; 1repeated ETA, 1 repeated ETA and tibialis anterior transfer (TAT), and 1 TAT),no one end with talectomy[14]. Boehm et al had 3 patients of 11 ended with relapse,2 patients were successfully treated with repeat casting and/or ETA; the remaining 2 relapsed clubfeet in one patient were treated with extensive soft-tissue-release operations[12]. Kowalczyk and Lejman had 7 feet with clinically satisfactory results and 3 unsatisfactory feet, there were 2 (one child) with rocker-bottom pseudocorrections after repeated bilateralETA and one recurrent clubfoot (one child). Six feet required soft tissue releases at 3, 12 and 21 months after the AT due to recurrence of moderate equinus and adductus. Three feet (two children) underwent repeat ETA at 10 and 15 months after the primary procedure [13].With van bosse et al, 4 patients (eight of 19 feet) had

early recurrences and required additional series of casts, 2 of which (4 feet) required one additional series and the other 2patients (4 feet) required two additional series(58).Hosam E. Matar et al, 3 patients with bilateral severe deformities (6/17, 35.3 %) had failed Ponseti treatment, despite initial correction. All 3 patients had multiple relapses, eventually requiring Ilizarov external fixator techniques,and two had persistent deformities requiring formal posteromedial soft tissue releases(59).In our cases Talectomy accounted for half of the relapsed cases (50.0%), while the remaining 50.0% were treated by repeated casting and heel cord release plus cast.

So we had satisfactory outcome in 12/16 (75%) patients after exclusion of patients end by talectomy, which was similar to Morcuende et al. in 2008 as they reported satisfactory outcome in 11/16 (67.75 %) patients(14).in 2016 Hosam E. Matar et al treated 10 patients with 17 syndromic clubfeet, they had a satisfactory outcome in 11/17 (64.7 %) patients(59).In 2008 Boehm et alhad a satisfactory outcome in 11/12 (92%) patients(12). Kowalczyk and Lejman also had satisfactory outcome in 7 feet (70%)(13).

Table 4.1: Summary of published studies on the use of the Ponseti method in treating clubfoot associated with arthrogryposis.

variables	Morcuende et al.(14)	Boehm et al. (12)	Kowalczyk and Lejman(13)	van Bosse et al.(58)	Hosam E. Matar et al(59)	Current study
No. of patients	16	12	5	10	10	16
No. of feet	32	24	10	19	17	30
Average follow-up, years	4.6	2	2.9	3	5.8	2.75
Satisfactory outcome	67.75 %	92 %	92%	78.9%	64.7%	75%

CONCLUSION:

The Ponseti method is effective treatment for syndromic clubfoot to achieve flexible, pain free and plantigrade foot, which historically has been difficult to treat nonoperatively, it is less extensive than previously report method of treatment and has minimal rate of complications.Our study support the use of Ponseti methods which is effective to get initial correction and to decrease the risk of extensive surgery in future. Even with relapses, has a role in treatment and minimizing the extensive surgeries.

Recommendation:

A long term follow up of those patients in order to base the evidence of these variable factors and to evaluate the outcomes and possibility of relapse.

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