Surgical Correction of Congenital Vertical Talus by One-stage Peritalar Reduction and Tibialis Anterior Transfer

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Abstract:

Twenty-eight feet of 16 children with true congenital vertical talus (CVT) were treated surgically by one-stage peritalar reduction and tibialis anterior transfer to the neck of the talus. The male to female ratio was 3:1. Fifty percent (8 patients with 15 feet) were isolated CVT and the remaining (8 patients with 13 feet) were CVT associated with other congenital anomalies mostly arthrogryposis. Patients with the neurological disorders were not included. Familial occurrence was encountered in 4 children (7 feet) of the same family. The mean age at operation was 19.1 ± 7.1 months (range 6-36).

All patients were operated by the author and were available for clinical and radiological follow-up for a mean of 9 ± 5.3 years (range 2-16).

A modified scoring system with 32 points for clinical and 8 points for radiological parameters was utilized for assessment of the final outcome.

Nine feet (32.1%) had excellent results, 15 (53.6%) good, 3 (10.7%) fair and one (3.6%) poor. The mean score for all patients was 31.9 ± 2.8 points. All patients and their parents, except one, were satisfied by their functional results and appearance of the feet.

Radiologically there was a statistically significant improvement of the measured angles at the final follow-up compared to the preoperative angles.

Unsatisfactory results occurred in 4 feet (14.3%); in 3 due to partial recurrence of the deformity; these were associated with arthrogryposis. The fourth had overcorrection. No talar avascular necrosis was encountered in this study.

Early treatment of CVT by simultaneous correction of all the anomalies during the same sitting led to very satisfactory medium term results.

Introduction:

Congenital vertical talus (CVT)[1-5] also known as congenital convex pes valgus[6-10] among other names,[11-17] is the most severe malformation of the spectrum of congenital flat feet.[2,5,20-24] It is a rare complex perplexing deformity with uncertain etiology,[1,7,18,25,26] and controversial methods of treatment.[4,7,8,10,18-20,27-52] It is characterized by its distinct clinical and radiological features, namely a rigid rocker-bottom deformity that produces "Persian slipper" configuration of the foot due to vertical placement of the talus and irreducible dorsal talonavicular dislocation.[1,2,5-7,11,13,15,25,26,31,53,54] (Fig. 1). It may be associated with a wide variety of disorders including neuromuscular, neural tube defects and other syndromes,[2,6,7,14,15,20,29,32,35,42,53-59] yet isolated cases may occur.[2,34,56] Familial occurrence has been reported,[6,31,34,56] and recent studies support an autosomal dominant transmission.[60]

The aim of the present study was to evaluate the results of surgical treatment of CVT by one-stage peritalar reduction and tibialis anterior transfer to the neck of the talus.

Subjects and Methods:

This study included 28 feet with true CVT in 16 patients; 12 (75%) were bilateral and 4 (25%) unilateral; both sides were equally affected. There were 12 boys (75%) and 4 girls (25%). Their age at operation ranged from 6 to 36 months with a mean of 19.1 ± 7.1 months.

The study was conducted at Al-Hadara Orthopedic University Hospital, Alexandria, during the period from 1985 to 1999 inclusive. All patients were seen, treated and followed-up by the author. Patients with neurological disorders (e.g. cerebral palsy, myelomeningocele, spinal dysmorphism, etc.) were excluded from this study.

The criteria for inclusion in the study were:

(a)- the diagnosis of true CVT with a maximum plantar flexion radiographs demonstrating a fixed and rigid abnormality of the talonavicular joint,[12,53]

(b)- A talar axis-first metatarsal base angle (TAMBA) of Hamanishi[56] above 60° that reflects the amount of dorsal dislocation of the navicular over the talus to differentiate true CVT from oblique talus (OT).

These patients were subdivided into two groups:

Group I: Isolated CVT i.e. not associated with other congenital malformation in the limb affected or elsewhere. These were 8 patients (50%); 7 had bilateral affection and one was unilateral, thus making a total of 15 feet.

Group II: CVT associated with other congenital deformities. These were 8 patients (50%); 5 had bilateral affection and 3 were unilateral, thus making a total of 13 feet. The associated congenital deformities were mainly the result of varying degrees of arthrogryposis without neurological deficits. They were as follows:

A. Unilateral cases (3): A girl with talipes equinovarus (TEV) of the contralateral foot; two boys with developmental dysplasia of the hip (DDH), extension contracture of the knees accompanied with TEV of the contralateral side in a boy and ptosis in the other one.

B. Bilateral cases (5): Bilateral flexion of the wrists, pollex varus and congenital hernia in a body. Idiopathic scoliosis and pelvic tilt in another body. Varying degrees of extension contracture of the knees in 3 patients, accompanied by congenital hernia in a boy (Fig. 2) and hemangioma of the face in a girl.

Familial occurrence was encountered in 4 patients (25%), 2 boys and 2 girls with 7 feet (25%) of the same family. Three of them had associated deformities (group II) and one was isolated CVT (group I).

All the patients were examined clinically and radiologically to assess the presence and severity of the deformity. Photographs were done for all the patients initially and at the follow-up examinations.

Radiological examination:

Conventional anteroposterior, lateral and lateral views with maximum plantar flexion, dorsiflexion and standing views were done. The following angels were measured.

A. Anteroposterior view angles:

1. Talocalcaneal angle (TC). It reflects the alignment of the hind part of the foot; normal range is 20^{o}-50^{o}.[61]
2. Talo-first metatarsal angle (TM1). It reflects the relation between the forefoot and hindfoot; normal range is -10^{o}-30^{o}.[61]

B. Lateral view angles:

1. Talocalcaneal angle (TC). it reflects the alignment of the hindfoot; normal range is 15^{o}-55^{o}.[61]
2. Tibiotalar angle (TiT). It is a measure of the position of the talus i.e. verticality of the talus; normal range is 85^{o}-145^{o}.[61]
3. Tibiocalcaneal angle (TiC). It is a measure of the equinus or plantar flexion of the hindfoot; normal range is 55^{o}-95^{o}.[61]
4. Talo-first metatarsal angle (TM1). It measures the relation between the hind and forefoot on the lateral projection; normal range is -5^{0}-40^{o}.[61]
5. Talar axis-first metatarsal base angle (TAMBA).[56] It measures the dislocation of the talonavicular joint; normal range ($<10^{o}$); good ($<30^{o}$); subluxated if TAMBA is between 30^{o}-60^{o} and dislocated (i.e. true CVT) when it is $>60^{o}$.

Methods of treatment:

A. Conservative treatment:

This was tried as a preliminary preparation for surgery when the child presented early aiming at stretching the skin, fibrous tissue structures and tendons of the anterior and lateral aspects of the foot and ankle. Gentle manipulation included stretching the forefoot into plantar flexion, inversion and adduction. The hindfoot was then manipulated by pushing the calcaneus upward and the heel downwards with inversion. The foot was fixed in the most corrected position. Manipulation and casting was repeated every 2-3 weeks for a maximum of 6 months.

B. Surgical treatment:

Under general anesthesia and tourniquet, the foot was approached through two incisions.

The first was a posterolateral one starting from the sinus tarsi and extending obliquely proximally below the lateral malleolus and then along the lateral border of the Achilles tendon (Fig. 3.A). Through this incision Z plasty of Achilles tendon with the distal cut directed laterally, posterior capsulotomy of the ankle and subtalar joints and release of the contracted fibulocalcaneal ligament were performed. The calcaneocuboid joint was identified and release of all tight structures around it was done, including the calcaneocuboid ligament and the bifurcate ligament, to correct the lateral column alignment. The peroneal tendons were lengthened by Z plasty if needed as well as the long toe extensor tendons and peroneus tertius. The dorsal and lateral talonavicular capsule was divided. This allowed the navicular to be more readily reduced over the talar head. In severe cases the lateral interosseous subtalar ligament might require partial or full release to correct the valgoid hindfoot so that the talus could be easily manipulated into position by means of blunt instrument.

The second incision was a medial one, curved over the talonavicular joint and centered over the talar prominence (Fig. 3.B-D). The navicular bone was riding over the anterior talar neck. Through both incisions, more release of the talonavicular joint

capsule and mobilization could be achieved. The midfoot could now be manipulated into plantar flexion with reduction of both the talonavicular and calcaneocuboid joints. After full release, the talus was elevated and forefoot manipulated.

The tibialis anterior tendon was released from its insertion, dissected proximally, transferred to the talar neck through a drill hole and sewing it to itself as a dynamic sling to prevent abnormal plantar flexion of the talus.

Careful capsuloplasty was important for maintaining the relation of the talus to the navicular, especially the plantar calcaneonavicular (spring ligament), which was sutured under tension. Maintenance of reduction of the talonavicular and calcaneocuboid joints was effected by drilling two Kirschner wires (K-wires) through them. Occasionally, a third one was drilled through the calcaneum to maintain the corrected heel position.

The lengthened tendons were sutured and the wound closed; lateral side first to avoid tension on the sutures (Fig 3. E-H). A well padded cast was applied in slight varus and equinus of the forefoot.

Post-operative regimen:

At two weeks, stitches were removed, the cast was changed into a moulded one. Wires were removed after 6 weeks. A short leg cast was applied for another 6 weeks. The foot was then supported by an ankle-foot orthosis with high medial arch for another 6 months. Subsequently ordinary foot-wear was used.

Follow-up:

The follow-up ranged from 2- 16 years with a mean of 9 ± 5.3 years. The mean age at final follow-up was 10.7 ± 5.4 years (range 2.5- 17.7 years).

Methods of assessment of the results:

A modified point-scoring system was applied taking into consideration previous reports on the clinical,[18,19,32,35,40,49] and radiological examinations.[35,36,46-49,51,52,56]

The modified scoring system used by the author incorporated 32 points for clinical parameters and 8 points for radiological parameters making a total score of 40 points (table I). The resulting total score at the end of the follow-up reflected the quality of the result and was classified as excellent (34-40 points), good (30-33 points), fair (26-29 points), and poor (25 points or less).

Statistical analysis:

Statistical analysis was performed with the use of Statistical Program for Social Science "SPSS" program on IMB compatible computer. The following tests were used to describe the collected data and analyze the obtained results: Student t-test, Chi-square test, Fischer exact test and correlation coefficient. The level of significance selected for this study was at $P<0.05$.

Table I: The point scoring system.

Parameter	4 points	3 points	2 points	1 point
I. Clinical (32)				
A. Appearance (16)				
Heel posture	Neutral	Mild valgus	Moderate valgus	Gross valgus or varus
Lateral border	Convex	Straight	Slight concavity	Gross forefoot abduction
Talar prominence	None	Minimal	Moderate	Callus or ulceration
Medial longitudinal arch	Normal	Decreased	Abscent	Reversed
B. Mobility (8)				
Ankle joint (4)				
passive dorsiflexion	Above square	To square	Slight equinus	Gross equinus
passive plantar flexion	>20°	10°- 20°	<10°	Fixed dorsi-flexion
Tarsal joints (4)				
Subtalar joint and Mid tarsal joint	Useful range of all tarsal joints	Subtal joint stiff other tarsal joints mobile	Stiffness of the whole tarsus	Rigidity of the whole tarsus
C. Symptoms (8)				
Function (4)	Normal	Mild reduction	Unable to do heavy work	Difficult walking
Pain (4)	No	Occasional	Painful after strenuous activity	Persistent pain
II. Radiological (8)				
1. Hindfoot correction (2 points), 2. Verticality of the talus (2 points), 3. Forefoot correction (2 points), 4.Dislocation of the talonavicular joint (2 points)				

Table II: Comparison between the preoperative and postoperative angles.

Angle	Preoperative			Postoperative			t	P
	Range	*Mean*	*SD*	*Range*	*Mean*	*SD*		
AP TC	30-50	41.43	5.25	15-45	25.54	8.09	11.88	0.006*
AP TM1	10-45	31.61	8.61	0-40	13.39	10.10	11.61	0.002*
Lat TC	35-52	44.21	3.82	20-45	35.00	6.25	9.17	0.004*
Lat TiT	130-180	159.68	12.77	80-150	99.86	16.41	18.86	0.007*
Lat TiC	100-135	118.11	10.13	70-130	87.68	13.3	21.71	0.002*
Lat TM1	35-90	60.18	16.47	-15-30	7.21	14.55	14.42	0.003*
TAMBA	60-90	73.64	7.36	-10-45	19.93	13.62	21.66	0.008*

AP= anteroposterior, Lat= lateral, TC= talocalcaneal, TM1=talo-first-metatarsal, TiT= tibiotalar, TiC-tibiocalcaneal, TAMBA= talar axis-first metatarsal base

** = Significant*

Results:

Using the modified point scoring system described before, the overall results at the end of follow-up were excellent in 9 feet (32.1%), good in 15 (53.6%), fair in 3 (10.7%) and poor in one foot (3.6%). Therefore, satisfactory results were obtained in 24 feet (85.7%) and unsatisfactory results in 4 (14.3%). The mean score at the end of follow-up for all patients was 31.9 $\pm$ 2.8 (range from 25-36 points).

Clinical results:

The appearance of 27 feet (96.4%) was cosmetically acceptable. They were plantigrade with well formed longitudinal arch (Fig. 4). However, some flattening was noticed in 8 feet (28.6%) on weight bearing (Fig. 5). A reversed arch with recurrence of rocker-bottom deformity was observed in one foot (3.6%) with callosities beneath the metatarsal heads and the depressed anterior aspect of the calcaneum.

The heel posture, viewed from behind during standing, was neutral in 18 feet (64.3%), mild to moderate valgus was noted in 9 feet (32.1%) (Fig. 6) while varus of the heel occurred in the remaining foot.

Residual abduction of the forefoot was observed in 6 feet (21.4%) with slight concavity of the lateral border (Fig. 5). Minimal medial talar prominence was noted in 4 feet (14.3%) with neither callosities nor ulceration.

The ankle movements were mildly restricted in 10 feet (35.7%). No foot with gross equinus or fixed dorsiflexion was observed. Slight equinus occurred in 2 feet (7.1%). The range of passive plantar flexion ranged from 10^o- 25^o with a mean of $18^o \pm 6.5$. Eighteen feet (64.3%) had some restriction of inversion and eversion varying between 25%- 75%. Rigidity of whole tarsus occurred in one foot (3.6%).

There was no functional limitations with painless feet in 20 (71.4%), mild reduction of activity and occasional pain in 5 (17.9%), painful feet after strenuous activity in 2 (7.1%) and difficult walking in one (3.6%).

Ordinary footwear was used in all except two feet (7.1%) which needed special shoe wear.

Radiological results:

Preoperatively, the mean value for the talocalcaneal angles were within the normal range according to Vanderwilde et al,[61] whereas the other angles were above the normal.

Postoperatively, there was a significant decrease of all angles ($P<0.05$). The talus became more horizontal (Lat TiT angle), the fixed equinus position of the hindfoot corrected (Lat TiC and Lat TC angles), the valgus of the hindfoot improved (AP TC angle), the abduction of the forefoot corrected (AP TM1 angle), the rocker-bottom deformity corrected (Lat TM1 angle) and the relationship of the talonavicular joint improved (TAMBA). Improvement in the measured angles, postoperatively, significantly affected the final score ($P<0.05$). This denoted the matching of the clinical and the radiological findings (table II).

There was no case of talar avascular necrosis in this study. On the other hand, some irregularities of the talonavicular joint was noted in some cases (Fig. 5. G-H).

Factors affecting the final score:

1. Age at operation:

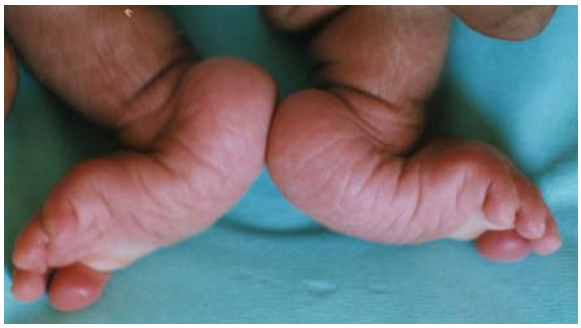
There was a trend that the younger the age at operation the better the score was. The best score was obtained in children operated upon during the first year (score= 33.2 $\pm$ 3.8). However, age has statistically insignificant influence on the final score in general ($P=0.47$).

2. Sex:

The mean score for females (33.0 $\pm$ 1.8) was better than that of males (31.0 $\pm$ 3.1). This difference was statistically insignificant ($P=0.16$).

3. Side and bilaterality:

There was no statistical significant difference in the mean score between the right and left sided affection (31.8 $\pm$ 3.5 and 32.1 $\pm$ 2.8 respectively; $P=0.17$). Also, the difference in the mean score between unilateral (31.8 $\pm$ 3.8) and bilateral cases (32.0 $\pm$ 2.8) was statistically insignificant ($P=0.52$).



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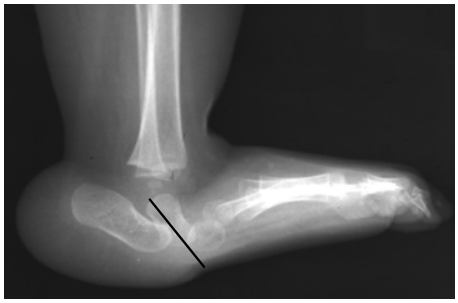

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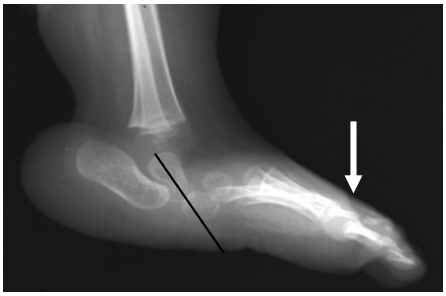

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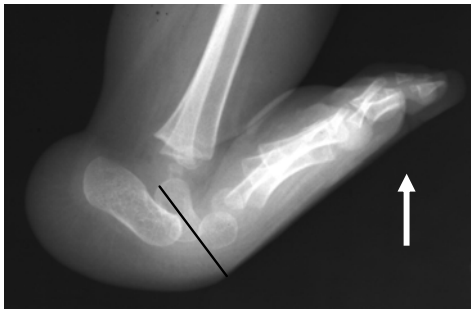

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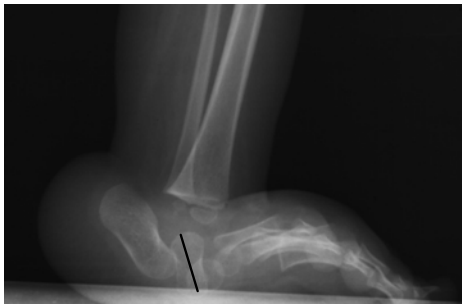


(E)

Fig. 1. Characteristic clinical and radiological features of true congenital vertical talus.

A. Bilateral CVT. Note the equines and valgus posture of the heel, the dorsiflexed, abducted, everted forefoot, the convex bulge of the sole and the upward concavity of the lateral border creating a typical rocker-bottom deformity or "Persian slipper" configuration of the foot.

B-E. Lateral x-rays of the foot with CVT, showing constant vertical orientation of the talus with irreducible talonavicular dislocation in plantar flexion (Fig. 1.C), dorsiflexion (Fig. 1.D) and standing views (Fig. 1.E).

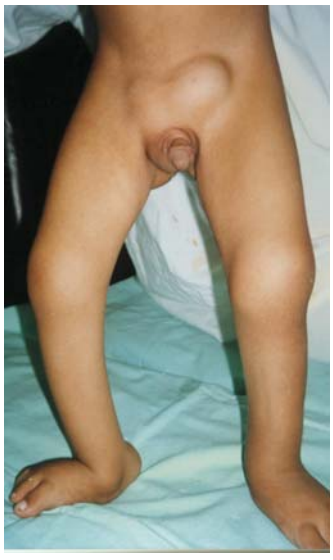


Fig. 2. Bilateral CVT associated with bilateral extension contracture of the knees and congenital hernia.

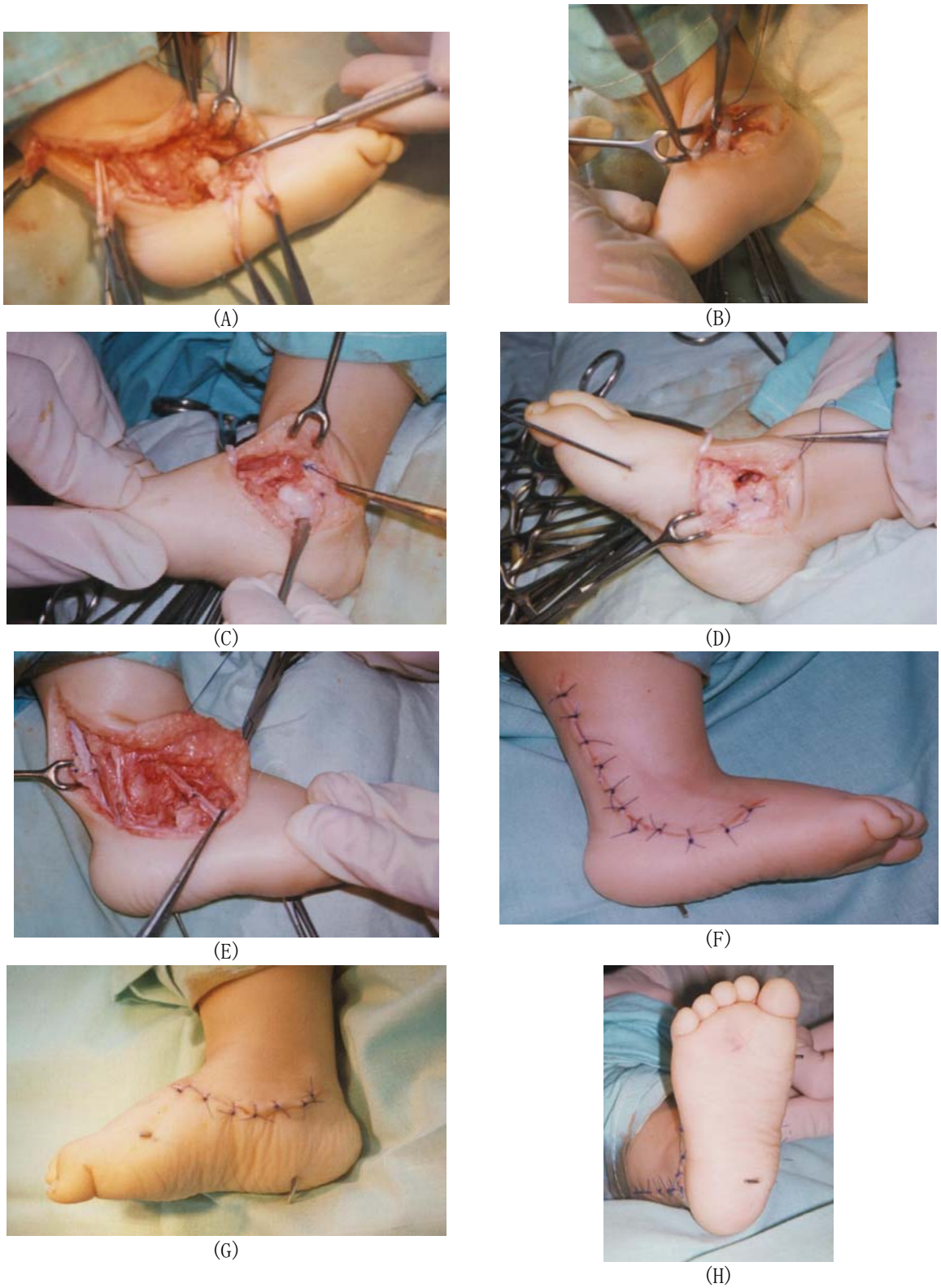


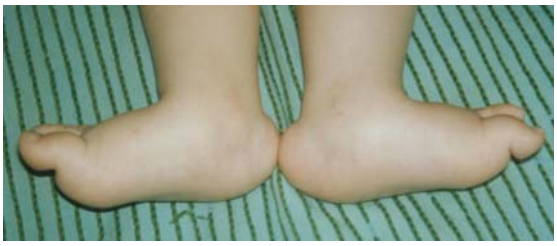
Fig. 3. Operative Procedure

A. *Posterolateral incision*. Z-plasty of Achilles' tendon, peronei and toe extensors. Dorsal and lateral talonavicular capsular release.

B-D *Medial incision.* Medial exposure of tibialis anterior tendon, more release of the talonavicular joint capsule, transfer of tibialis anterior tendon to the talar neck, plication of spring ligament, elevation of the talus, fixation of the reduced talonvavicular joint by K-wire.

E. Repair of the lengthened tendons.

F-H. Appearance of the foot after closure of the wounds and K-wires fixation.



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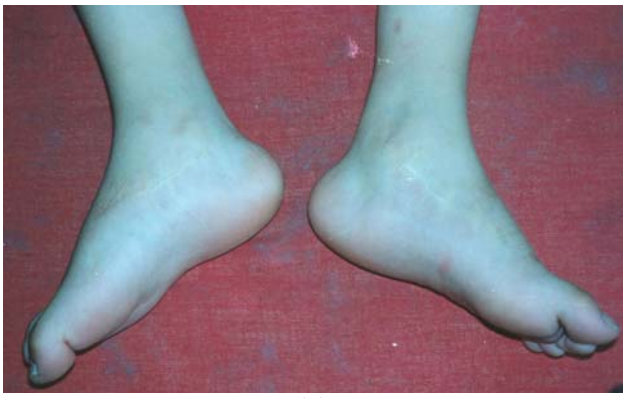

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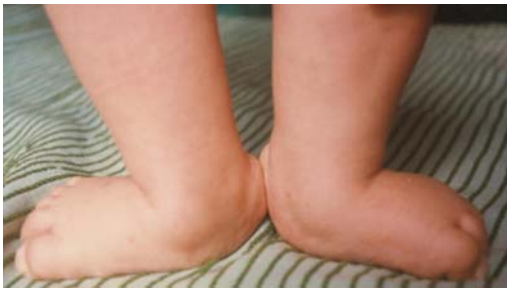

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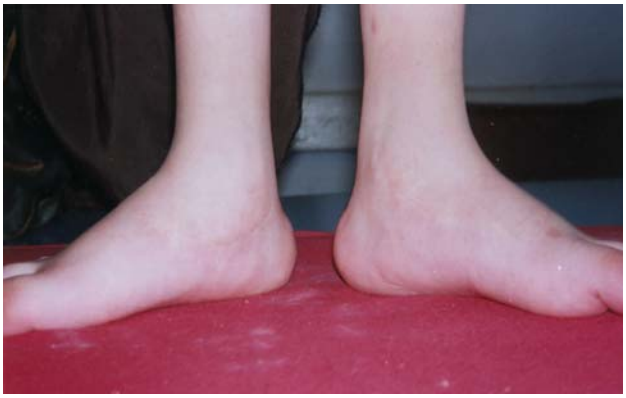

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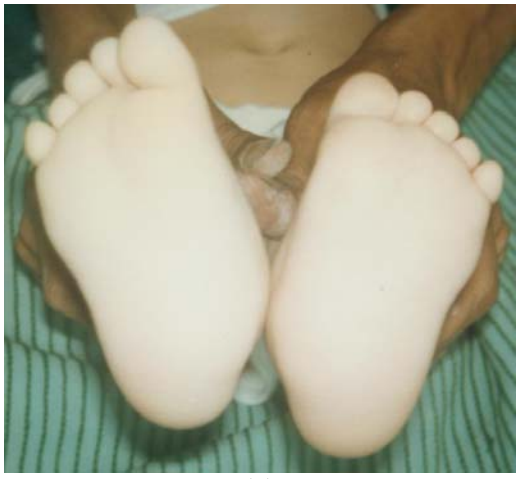

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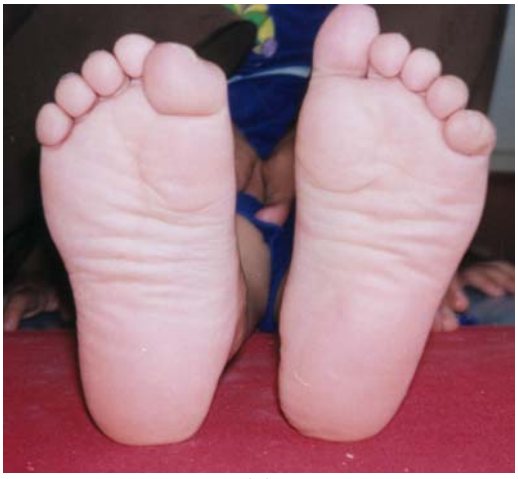

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Fig. 4. Bilateral CVT in a boy of the isolated group
(Clinical photographs of the same case in Fig. 3).

A-C. Preoperative photographs (age of 7 months).
D-F. Follow-up photographs (age 6 years)

Note:
- Plantigrade feet with well formed medial longitudinal arch.
- The corrected inferomedial talar protrusion. Satisfactory results.

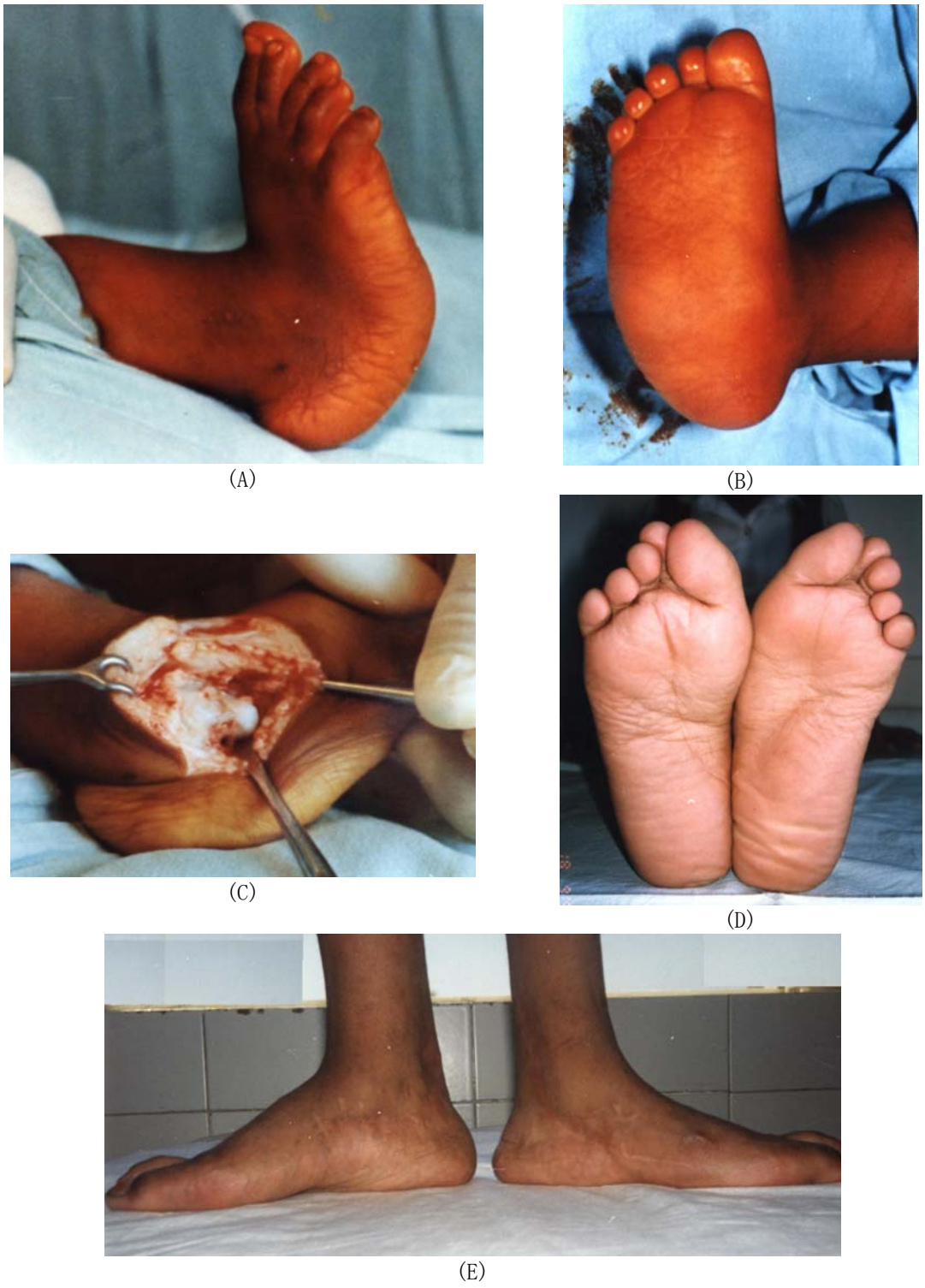
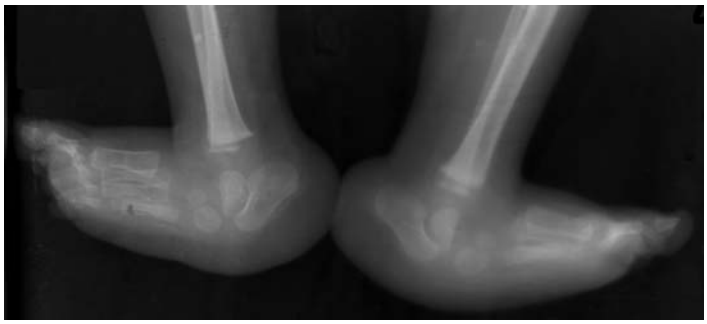


Fig. 5. Bilateral CVT in a girl of the associated group

A&B. Preoperative photographs of the right foot at the age of 2 years showing the rocker-bottom deformity and the talar prominence.
C. Vertical orientation of the talus intraoperatively
D&E. Follow-up photographs at the age of 17 years showing the plantigrade feet, well formed arch of the right foot, flattening of the left foot on weight bearing with residual abduction of the forefoot.



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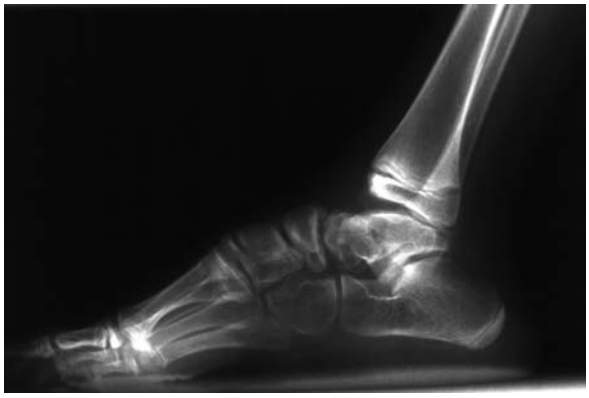

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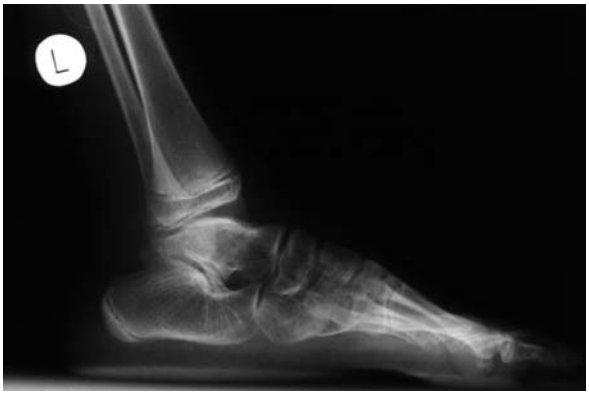
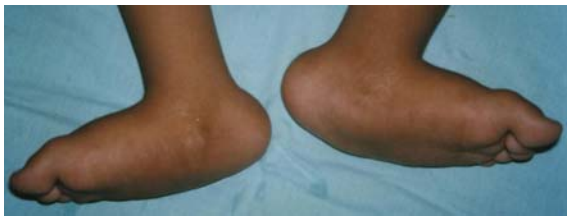


(H)

Fig. 5. (continued)

F. Preoperative x-ray.

G&H. Standing x-rays at the end of follow-up showing horizontal orientation of the talus, well formed arch on the right side, flattening on the left side and irregularities of the talonavicular joint.



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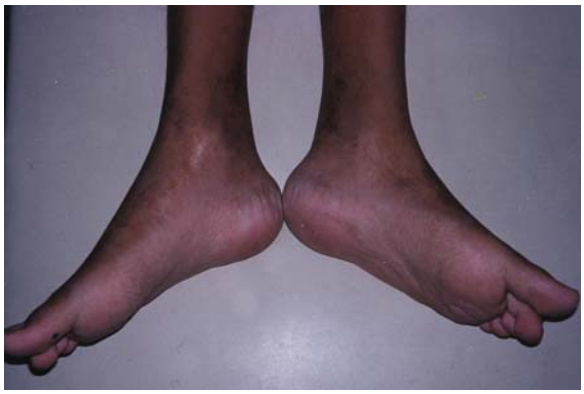

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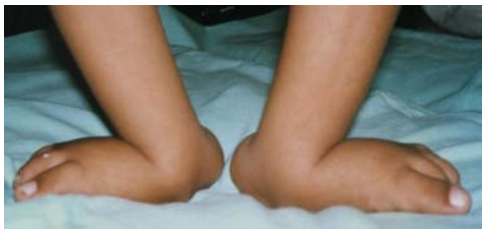

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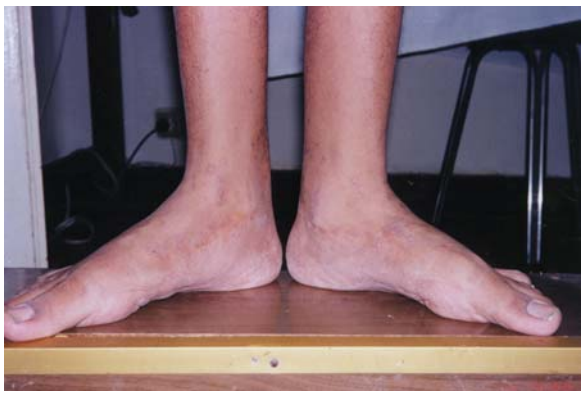
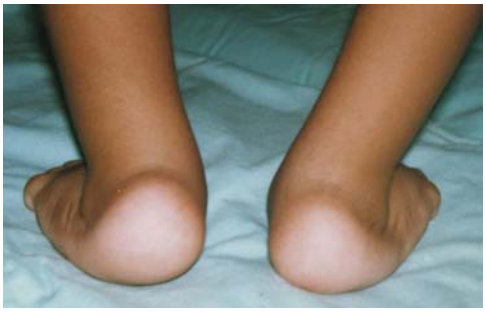
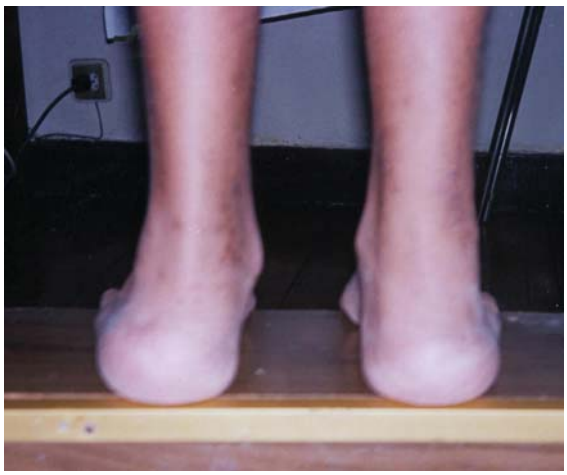


(E)



(C)



(F)

Fig. 6. Bilateral CVT in a boy of the associated group

A-C. Preoperative photographs at the age of 1 year.
D-F. Follow-up photographs at the age of 16 years.

Note: Plantigrade feet with well formed arch, residual valgus of the heels on weight bearing.

4. Length of follow-up:

There was a negative correlation between the period of follow-up and the final score. Half the cases were followed for more than 10 years (mean = 13.9 ± 1.1 years), with a mean score of 31.4 ± 3.3 compared to a mean score of 32.5 ± 2.2 for cases followed for less than 10 years. However this difference was statistically insignificant (P=0.13).

5. Etiological groups:

The mean score for group I (32.5 ± 0.3) was better than group II (31.3 ± 3.2). This difference was statistically insignificant (P=0.29).

6. Familial occurrence:

The mean score for familial cases (30.7 ± 3.8) was less than the other cases (34.0 ± 2.8). This difference was statistically insignificant (P=0.06).

Analysis of unsatisfactory results:

Unsatisfactory results were seen in 4 feet (14.3%) of three males (19% of the males), three feet were of bilateral affection (12.5% of bilateral cases), three were on the right side (21.4%), three were of the associated group (23.1%) and three of familial occurrence (42.9%).

It was observed that unsatisfactory results in cases operated below 2 years were 13.0% (3 fair cases out of 23) while it was 20.0% (one poor out of 5 cases) among cases operated above 2 years.

There was a statistically significant difference (P=0.04) between the mean age at final follow-up for satisfactory and unsatisfactory results (9.8 ± 5.4 and 15.8 ± 1.4 years respectively). Also, the mean length of follow-up for unsatisfactory results (14.2 ± 1.5 years) was more than that for satisfactory results (8.2 ± 5.2 years) and this difference was statistically significant (P=0.03).

There was no statistically significant difference in the measured preoperative angles between the unsatisfactory and satisfactory results in spite of being higher in the former (P>0.05). On the other hand, improvement in the measured angles at final follow-up was more obvious in the satisfactory results and this was statistically significant (P<0.05) with exception of the anteroposterior talocalcaneal and the lateral talo-first metatarsal angles which although improved yet the difference was statistically insignificant (P=0.67 and 0.34 respectively).

The worst results were obtained in 2 patients with arthrogryposis (3 feet) due to some recurrence of the deformity in spite of good early postoperative correction. The fourth foot of a bilateral CVT child with no other deformities was due to overcorrection with cavovarus deformity developed five years postoperatively.

Discussion:

Congenital vertical talus is a rare enigmatic pediatric foot deformity characterized by its resistance to conservative and operative treatment and the cosmetic and functional blemishes it causes at the time of adolescence is reached.[1,2,4,5,7,18,20,62] It is essential to recognize that CVT does not delay walking, and if delay is noted an underlying cause must be sought before treatment commences.[10,20,21,63] Untreated cases result in awkward clumsy gait with deterioration of the shoe wear.[1,2,8,18,20,21,42,52]

The exact incidence of CVT is unknown. Osmond-Clarke[1] encountered the deformity once in 131 consecutive cases of congenital foot deformities of which 121 were congenital talipes equinovarus. It is estimated to be one for every 10000 live births.[9,47]

Boys are more commonly affected than girls,[1,4,6,10,19,29,42,45,48,55,56] with a ratio of 3:1 in the present series. Bilateral involvement occurred in 50% [20,47] to 71% [42] of reported cases and was 75% in the present study.

Lloyd-Roberts and Spence [2] distinguished four clinical forms according to whether the deformity was isolated or not. Ogata et al.[34] proposed a classification system that divides patients into one of the three groups based on whether the condition is primary isolated form with no other congenital abnormalities, associated with other congenital abnormalities and without neurological deficits and those associated with neurological deficits (paralytic vertical talus). Hamanishi [56] classified his cases into five groups and included those with chromosomal aberration in his classification. Lichtblau [64] defined three groups; teratogenic with positive family history, neurogenic associated with muscle imbalance and acquired attributed to malpositions. The incidence of CVT in patients with myelomeningocele who had foot deformities had been found to be 10%. [55]

In the present series, patients with neurological disorders were excluded. These patients have different prognosis and muscle-balancing procedures may be required as part of the operative correction.[20,55,65]

The primary isolated form (group I) was found in 50% of our patients. A nearly similar ratio was noted by previous authors.[2,32,34,47]

Although different forms and degrees of CVT were identified, most authors still approach these patients clinically as a single anatomopathologic entity.[2,23,34,47,52,56] In the present study, there was no difference between these groups clinically or radiologically as far as the feet are concerned. Though there is a constant dislocation of the talonavicular joint there are varying degrees of severity of all the

elements of deformity.[13] Coleman and his colleagues [29] distinguished two types of CVT; those with talonavicular dislocation and the second with concomitant dislocation of the cuboid over the calcaneum.

Patients who have arthrogryposis have fixed vertical tali as part of their clinical spectrum.[56,57,66,67] These forms of vertical tali tend to be more rigid and have less favorable outcome than those associated with other congenital malformations.[20] This was evident in the present study as 75% of the unsatisfactory results were among patients with varying degrees of arthrogryposis.

Familial occurrence of CVT was first reported by Lamy and Weissman [6] in each pair of homozygous twins, in two brothers and also in a father and his son. They suggested a hereditary factor. Robbins [31] reported occurrence in a mother and daughter. Ogata et al.[34] found familial incidence in two families; mother, son and daughter in one family and 2 siblings (sister and brother) were involved in another family. They also reported 8 other cases whose first degree relatives were reversely affected (club foot, pes cavus and metatarsus adducts). Many associations of CVT and oblique talus (OT) in some patients in the same family have been observed by Hamanishi [56] and in this work too.

In the present series, familial occurrence was encountered in 4 children with 7 feet; a boy and his sister, their mother has bilateral CVT; a girl, her father has unilateral CVT and OT on the other side; a boy with unilateral affection CVT with TEV and DDH on the contralateral side, his father also had bilateral feet affection. The two fathers and the mother of these children were siblings. Their grandfather had also bilateral feet deformity, the nature of which was not known. Genetic factors may play an important role in the etiology of primary isolated forms. Stern[68] described a family with autosomal dominant pattern but this is unusual.

The objectives of treatment of CVT include restoration of the normal anatomical relationship among the talus, navicular, calcaneus and cuboid and maintaining reduction so that a plantigrade, weight bearing surface within the sole of the foot could be provided in order to correct awkward gait and improve shoe wear.[7,16,19,20,50] It is generally accepted that conservative treatment is usually unsuccessful in the treatment of CVT and consequently surgical reduction is always necessary.[1-3,7,16,19,28,29,33-35,42,43]

The orthopedic manipulative treatment is difficult as we have to do two opposed actions, lower the forefoot and raise the hindfoot. It is the rigidity of the foot in true CVT which is responsible for its notorious lack of response to manipulative treatment.[1,2,7,53] It is probable that anatomic forms that respond to such treatment are in fact benign forms which correspond to congenital flat foot.[4,21,38,47] Vigorous manipulative efforts during infancy and childhood are not advised since they are ineffective and the damage produced is more notorious than if the condition was left without treatment.[13]

In the present work, as well as others,[4,14,16,28,30,47,51] manipulative treatment started early was at best an adjuvant to the surgical treatment as it rendered the feet more supple and facilitated the surgical correction.

Surgery remains the mainstay for treatment, however, the timing and precise procedure required to correct the deformity are contentious. The difficulty of surgical correction depends on the severity of deformity, the associated anomalies and the age of the patient.[1-5,18,29,30,42,45]

In the past, CVT was treated lately. This explains the frequency of osseous resections in old surgical techniques as talectomy,[6] naviculectomy[2,7,18,27,31,32,53] and also subtalar arthrodesis[3,8,19,29,34,43,46] and even triple arthrodesis. [7,13,16,19]

Most authors recommend open reduction in CVT at the earliest possible time. As with many juvenile foot deformities, the earlier and more efficiently the problem is tackled the better and more favorable prognosis will be.[41,68] Early treatment allows for bringing these developing structures into optimum use.[69] It leads to satisfactory results usually avoiding extensive procedures including tendon transfer and lengthening.[41,45] The sooner the deformity is corrected fully the less remodeling of the talus is needed.[33] Patterson et al.[25] showed that the talus is already deformed at birth and early full reduction may facilitate remodeling. Surgical correction of CVT is not difficult below 2 years but becomes increasingly formidable in later years because of secondary adaptive changes.[41,56,70] Some authors even recommended to operate at the age of 3-6 months.[33,45]

In the present study, the mean age at operation was 19.1 $\pm$ 7.1 months. Twenty three feet (82.1%) were operated below 2 years. The best score was obtained in feet operated during the first year (33.2 $\pm$ 3.8) and there was a trend the younger the age the better was the final score. Unsatisfactory results in cases operated below 2 years were 13% compared to 20% for cases above 2 years. In Duncan and Fixen[10] series, the average age of their patient was 31 months. They elected to postpone surgery until the child could walk in those patients with developmental delay. In some of their patients and in the present work too, the delay was due to late referral. Napiontek[46] observed poor radiological results in children operated after the fourth year of age. In older

children, the main problem is compounded by unchecked growth of the talar head and neck which makes the medial longitudinal column excessively long.[20] This deformity may require excision of the navicular to equalize the length of the medial and lateral columns[32] or even subtalar extra-articular Grice arthrodesis.[29,43,46]

Understanding the mechanism of production of the deformity leads to logical approach to its correction.[41] The key features of the pathoanatomy were clearly established.[25,26,70,71] Since the main pathoanatomy is the contracted dorsal tendons and the dislocation of the talonavicular and occasionally calcaneocuboid joints, all authors treating CVT deal with these structures either in two stages [3,7,11,29,40] or more recently in one stage.[10,34,39,41-52] Extensive capsular releases and tendon elongation performed have been described, some including capsuloplasty of the calcaneonavicular ligament.[16,29,33,38,40,52] Meticulous surgical technique is essential to obtain the best results in CVT.[8] The technique adopted in the present study addresses all the components of deformity in one stage. In systemic step-by-step approach the obstacles of reduction including the tight tendons and the contracted soft tissue i.e. ligaments and capsules were dealt with.

The talocalcaneonavicular joint bears special importance in CVT. Because of its morphologic similarities, this joint is also called coxa pedis[58] or acetabulum pedis[72] and as in the hip joint, dysplasia can occur. Defective development of the articular surfaces may lead to alterations in this joint such a talar protrusion or subluxation which is one of the clinical features in CVT. This joint must be reduced and reduction maintained appropriately in order to achieve satisfactory results.[52] We agree with other authors[10,20,39] that if full release of the talocalcaneonavicular joint is performed from the medial and lateral sides, naviculectomy will not be necessary as this may destroy the integrity of the medial ray.[40]

The release of the subtalar joint is a matter of controversy. Some authors[1,20,46,52] routinely release it, others[19,29,47] occasionally dived the lateral part of the talocalcaneal-interosseous ligament so that the talus can be easily maneuvered into position by blunt instrument as in the present work. The opponents of the release found that it was not required to achieve reduction and feared that this release may compromise the blood supply of the talus,[10,40] which fortunately did not occur in the present series.

In order to correct the lateral column shorting capsulotomy of the calcaneocuboid joint was done in all cases in the present series, and was recommended by others.[20,29,47,50,52] Seringe et al.[71] prefer to do dorsal wedge osteotomy of the large apoplysis of the calcaneum rather than opening the calcaneocuboid joint. In older children (5 years), elongation of the lateral column by interposing graft into the calcaneocuboid joint along with peritalar release and Grice arthrodesis was preferred by Marciniak.[73]

Primary tendon transfers were done especially in paralytic forms of CVT.[65,74] Osmond-Clarke[1] transferred the peroneus brevis to neck of the talus to act as an anchor to the talus in a more or less horizontal position and also as an active evertor of the talus. Coleman et al.[29] transferred tibialis posterior tendon to the plantar aspect of the navicular. However, the most popular transfer is that of tibialis anterior either split transfer or total transfer to the neck of the talus as advocated by Llyod-Roberts,[2] Stone,[27] Colton[18] and others[10,16,19,35,36,39.43,51,74] This transfer was used in all cases in the present series. The objectives were: to remove a deformity force causing dorsiflexion of the forefoot, to maintain the reduced coaxed talus with the navicular and first metatarsal and probably to act as dynamic transfer. Peroneus longus transfer to the talar neck as an alternative to absent tibialis anterior was used by Masterson et al with excellent correction in two cases with revision surgery.[75] On the other hand, open reduction was not accompanied by any transfer in some other reports.[33,41,45,48]

Many surgical incisions were utilized to achieve open reduction in CVT. They entail: staged multiple incisions,[7,29] single stage medial and posterior approach,[34] three incisions,[19] single stage dorsal approach,[41] transverse dorsal approach,[33] posterior dorsal approach[51] and medial approach.[44] Cincinnati incision that was originally proposed for surgical correction of CVT[76] was repopularised in recent studies.[20,47,48,50,52] The most important advantage of this incision is the possibility to reach, completely release and reduce the talocalcaneonavicular joint which is crucial for correction while its main handicap is the difficulty to reach the dorsal anatomic structures.[48,52]

Harrold[4,5] mentioned and Colton[18] agreed that CVT had no tendency to recur or relapse after adequate surgical correction. The extreme stability of the reduction thus obtained is reflected in the good results and emphasized by the disappointing outcome whenever one of the various steps has been omitted.[18] The commonest reason for surgical failure is inadequate reduction of the navicular.[13,18,46] Therefore, it is important to achieve full reduction of the navicular as part of the initial procedure.[13,26,50,52] Napiontek[46] reported the results of 23 children with 32 feet after 9 years follow-up. The extent of surgical release was tailored to the severity of deformity. He

observed that those with severe deformity and less extensive surgery had worse prognosis. It should be stressed that marked overcorrection of the talonavicular displacement should be obtained because postoperative X-ray may show that distortion of these bones deceived into incomplete reduction.[13] A common error is to take X-rays with the foot in plantar flexion which may be misleading.[19] Rarely, the foot function may be satisfactory without complete reduction.[7,16,32] However, incomplete reduction appeared to be compatible with satisfactory asymptomatic function but anatomic reduction is preferable.[13,32] On the other hand, Lichtblau[64] emphasized that early reduction can look good, but resubluxation is an insidious process and long term follow-up is needed. It is believed that this deterioration is due to plastic deformation of the talonavicular joint and until remodeling takes place resubluxation may occur.[52] Deterioration of correction has been reported by some authors[39,40,42,50-52] and observed in the present study in spite of perfect initial correction.

Accurate maintenance of reduction is essential by doing capsuloplasty, K-wires fixation or dynamic transfer. All these tools were used in this study. However, departure from any step of the technique may result in unsatisfactory outcome. K-wires problems as removal before anatomic and functional adaptation to the corrected position, cutting out through the poorly ossified navicular in young children or even when K-wires were omitted were among the technical errors that have been committed and reported.[13,18]

Unrecognized talonavicular coalition during correction of CVT may cause persistence or recurrence of the deformity. If the talus can not be adequately reduced at such operation or the valgus position of the calcaneum can not be corrected despite appropriate soft tissue release, the problem may be due to tarsal anomaly. An undetected coalition may lead to an incomplete or suprious reduction. Examination of the subtalar joint under direct vision at operation will prevent this error.[77]

Late results have not been as good as early results in the present study and some other studies too,[8,13,40] suggesting that not only because of the errors in treatment but also because of the underlying conditions mainly arthrogryposis, has added additional problems as the child grew older. Triple arthrodesis may be required as final procedure.

The mean period of follow-up for satisfactory cases were less than those for unsatisfactory cases (8.2 ± 5.2 and 14.2 ± 1.5) and this difference was statistically significant (P=0.03). This could be explained, among other factors mentioned, by improvement in the technique, being more meticulous and trying to avoid the errors committed in earlier cases.

Dodge et al.[42] have reported the largest series of patients with surgically treated CVT; a retrospective series of 36 feet in 21 patients with a mean follow-up of 14 years. All had variety of procedures performed by different surgeons. They were unable to show any differences between the groups of patients treated using alternative approaches.

Overcorrection after surgical correction of CVT was reported by Herndon and Heyman[7] in one case. They believe that overcorrection with the foot goining to varus was caused by lengthening of the peroneal and extensor tendons to such a degree that muscle imbalance developed and the invertors pulled the foot into varus. Wirth et al.[45] also reported overcorrection in one foot among 13 feet operated early between 3-6 months. These authors[7,45] did not transfer tibialis anterior tendon. Gradual development of cavovarus deformity in one foot after tibialis anterior transfer was reported by Duncan and Fixen[10] and by Kodros and Dias[50] also in 3 feet of two patients which required reoperation. They thought that the condition might be due to unopposed peroneus longus in its plantar flexion of the first ray after tibialis anterior transfer proximally. Zorer et al.[52] claimed that this deformity did not develop in their patients possibly because they have not transferred the tibialis anterior tendon but only lengthen it in 4 patients. Some of the Grice procedures performed by Napiontek[4,6] also resulted in overcorrection.

In the present study, overcorrection resulted in unsatisfactory fair result (score 28) in one foot of a boy with bilateral CVT of the isolated group that developed gradually five years postoperatively. He wears special shoe and he and his family were neither satisfied nor dissatisfied with the clinical outcome of this foot.

Avascular necrosis of the talus (AVN) has been described as complication of some of the procedures done for this deformity.[1,8,19,31,35,36,43,47] It is the result of extensive denudation of the talus during reduction of the dislocation[1,19] especially over the superior aspect of the talar neck,[7] forceful dorsiflexion of the navicular on the talus without adequate mobilization and failure to perform Achilles lengthening before or at time of surgery when performed in two stages.[8] Recently performing one stage open reduction decreased the incidence of AVN. In the present study and some other reports,[10,34,40,49,50,52] this major complication was not encountered. However, some irregularity of the neck of the talus may occur due to the presence of a tunnel for tendon transfer and due no doubt to vascular disturbance but the body of the talus was normal; this was observed by Osmond Clarke.[1] The irregularity of the talonavicular joint noted in some

cases in the present series and reported by other authors[10,41] may be due to the routine use of K-wire to fix this joint after reduction. Magnetic resonance imaging (MRI) provides a clear outline of the cartilaginous analage and possible avascular necrosis of the talus.[52] Unfortunately MRI studies were not available in our hospital at the time of the present study.

Many scoring systems were proposed in the literature for evaluation of the outcome of surgical treatment in CVT. Some, depended on the clinical aspect and morphological appearance only.[18,19] Others used both clinical and radiological criteria.[32,35,36,40,46-49,51,52,56] The modified point scoring system utilized in the present study is a more comprehensive one and incorporates the clinical and radiological parameters included in previous reports to substantiate with more accuracy the outcome of treatment after a mean period of follow-up of 9 ± 5.3 years. Although there was variable degrees of restriction of the ankle and subtalar joints as reported by other authors,[40-42,48,49,52] yet, pain and functional limitation were not an immediate or long term problem the present study. A divergence between relatively good appearance of the operated feet and worse radiological features has been observed by Napiontek.[46] On the contrary, the radiological results were superior than the morphological and clinical results in other reports.[47,78] In the present work both the clinical and radiological results were nearly matching. It appears that some of the improvement in the results and fewer complications may be seen from increased knowledge and understanding of the CVT pathoanatomy. Recurrence of deformity and talar avascular necrosis in the modern era, although worrisome are not so frequent.[51]

Conclusion:

The treatment of true CVT is surgical. The single-stage peritalar reduction and tibialis anterior transfer corrects the best observed lesions. The liberation may be gradual according to the severity of deformity. Early treatment promises the best results before adaptive changes occur. The procedure adopted in this work is suitable for both isolated and associated cases of CVT. It gives satisfactory results both clinically and radiologically. Poor prognostic signs include late age of surgical correction, laxity of meticulous technique and the association with arthrogryposis. Familial occurrence has been recorded in this study and genetic counseling is advisable. Since relapse of the initial correction may occur, a longer term follow-up is recommended.

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