CONGENITAL SYNDACTYLY: CASE BY CASE ANALYSIS OF 47 PATIENTS

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ABSTRACT

Objective: To assess and report clinical data from patients with syndactyly. Methods: A retrospective review of 47 patients treated between April 2002 and April 2012. Results: Among the 47 analyzed patients, 33 (70%) were male and 14 (30%) female. The total number of syndactylies was 116. The right hand was affected in 19 patients (40%), the left hand in 12 (24%) and 31 (36%) were bilaterally compromise. Sixteen patients (34%) also presented genetic syndromes. Among the 31 (66%) patients without syndromes, 12 (39%) had isolated syndactyly and 19 (61%) presented association with other hand anomalies. The third web space was affected 45 (39%) times; the fourth, 35 (30%) times; the second, 22 (19%) times and the first web space 14 (22%) times. Simple syndactyly was found 68 (59%) times, complete syndactyly in 44 (65%) and incomplete in 24 (55%). Complex syndactyly was found 48 (41%) times. Conclusion: The results in this study are similar to the literature. ***Epidemiological Study.***

Keywords: Hand deformities, congenital. Syndactyly. Child.

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INTRODUCTION

Syndactyly is a defect in the connection between two or more fingers. It is one of the most common congenital anomalies of the upper limb and occurs in approximately one in 2500 births.[1] It can occur isolated or associated with other malformations. Its cause is a failure in the differentiation of mesenchymal structures in single digits, where the longitudinal interdigital necrosis does not occur between the sixth and eighth week of intrauterine life. It is usually bilateral and symmetrical, mainly affecting males and is uncommon in blacks. It is most common between the third and fourth fingers, followed by the fourth and fifth and the second and third. Syndactyly between the first and second fingers is rare because the thumb of the hand is separated before the rest of fingers.[2]

The deformity is usually a result of sporadic mutations, but there are reports of autosomal dominant inheritance with variable expressivity in up to 40% of cases.[3] It is classified as simple when the fusion occurs only by the skin, being subdivided into complete or incomplete depending on the extent of interconnection, and as complex as the fusion occurs also through the bone structure of the fingers, with the possibility in such cases of abnormal tendineae, vasculonervous[2] and ungal.[4]

Several surgical techniques have been described to correct this deformity, most of them using skin grafts, since the surface area of the fingers apart is greater than the area of the fingers united.[5-9] There is also available the classic surgical technique associated with the patchwork degreasing with subsequent healing by secondary intention of bloody areas, in order to simplify the procedure and avoid possible complications due to grafts use.

MATERIALS AND METHODS

A retrospective descriptive study based on analysis of data from medical records of patients with syndactyly treated by Discipline of Hand and Microsurgery, *Hospital das Clínicas de Ribeirão Preto, Universidade de São Paulo*, Brazil over a 10 years period (between April 2002 and April 2012). Secondary cases due to trauma and burns were excluded, totalizing 47 patients with congenital syndactyly. Of these, we investigated the presence of genetic syndromes, other malformations, the total number of syndactylies, gender, affected side, the affected space, symmetry, whether simple or complex and whether partial or complete. Data were tabulated and expressed in absolute and relative frequencies. This study was submitted to and approved by the Institutional Ethics Committee Research.

RESULTS

Of the 47 patients with congenital syndactyly, 33 (70%) were male and 14 (30%) female. Sixteen (34%) patients had some genetic syndrome (six cases of Apert syndrome, two cases of Down's,

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one of Poland's and seven different syndromes), 31 (66%) patients had no associated syndromes. In 17 (36%) patients the disease was bilateral in 12 (70%) of these cases being symmetric. The right side was affected in 19 (40%) patients and the left side in 12 (24%). (Table 1) The total number of syndactylies was 116, and the third web space was affected 45 times (39%), followed by the fourth space 35 times (30%), the second space 22 times (19%) and the first space with 14 times (12%). Simple syndactylies accounted for 59% (68) of the total, 35% of them (24) were partial and 65% (44) complete. Complex syndactylies were 41% (48) of the cases. (Table 2, Figures 1 and 2)

Of the 31 (66%) patients without any associated syndrome, 12 (39%) had isolated syndactyly and 19 (61%) associated with other malformations. The most common malformation was acrosyndactyly in five cases; chipped hand in four patients; polissindactily, brachydactyly and several agenesis in three different situations each. Only one patient had associated congenital constriction band.

Table 1. Characteristics of patients with syndactyly.

	Total (n = 47)	%
Gender		
Masculine	33	70
Feminine	14	30
Associated Syndrome		
Yes	16	34
No	31	66
Affected Side		
Right	19	40
Left	12	24
Bilateral	17	36

Table 2. Characteristics of syndactylies.

	Total (n = 116)	%
Affected space		
1º space	14	12
2º space	22	19
3º space	45	39
4º space	35	30
Type		
Simple	68 (24 partial / 44 complete)	59 (35/65)
Complex	48	41

DISCUSSION

The importance of this type of case series is to compare the data in the literature with those found in practice at our service. The number of cases of congenital syndactyly in this study outperforms most publications related to this topic. Ekerot published in 1996 his experience with 11 patients and 17 syndactylies over a period of three years.[6] Withey *et al.*[10] presented their results on 19 syndactylies in 12 patients in 2001. Deunk *et al.*[4] reported in 2003 a case series of 27 patients over 21 years. Lumenta *et al.*[11] in 2010 published their experience on 26 syndactylies in 19 patients over a period of 42 years. Greuse *et al.*[12] in 2001 reported the evaluation of 16 patients presenting 24 syndactylies in two years. Works that include the highest number of patients are those of Bandoh *et al.*[8] in 1997 with 58 patients in nine years; D'Arcangelo *et al.*[9] in 1996 with 50 cases and 122 syndactylies in nine years, and the impressive study with 681 patients in 20 years in the study of Muzaffar *et al.*[13] published in 2004. In our midst Barboza *et al.*[14] in 2006 reported experience with 13 patients in two years, and Cortez *et al.*[15] with 72 patients in five years in 2010. (Table 3)

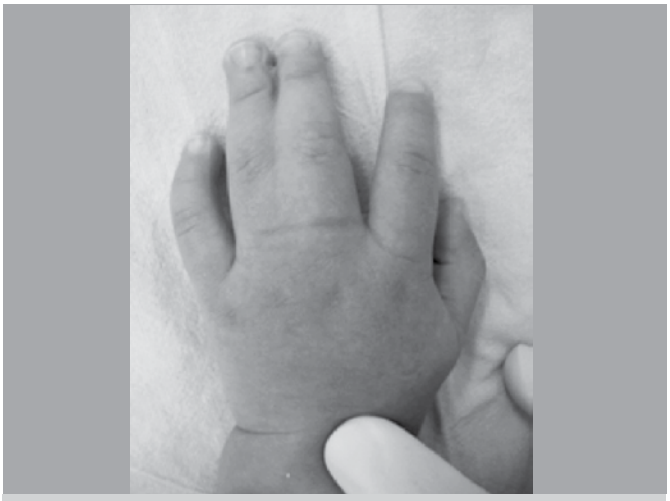

Figure 1. Complete syndactyly between the third and fourth fingers (third space) in a 1 year 5 months old child (dorsal aspect).

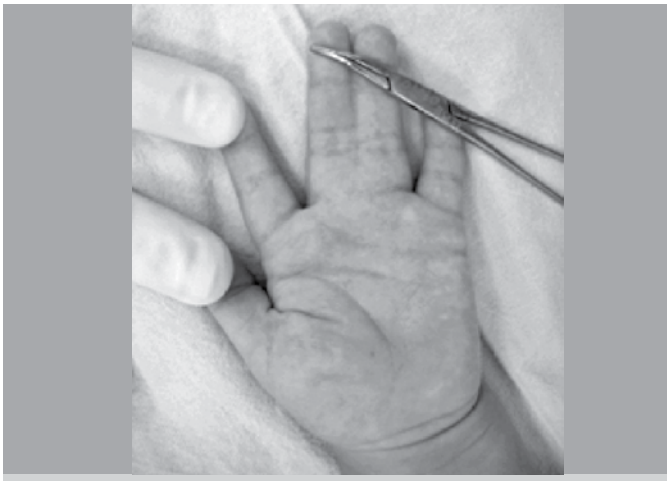

Figure 2. Complete syndactyly between the third and fourth fingers (third space) in a 1 year 5 months old child (palmar aspect).

Table 3. Recently published case series.

Publication year	Author	Number of cases	Period (years)
1996	Ekerot[6]	11	3
1996	D'Arcangelo *et al.*[9]	50	9
1997	Bandoh *et al.*[8]	58	9
2001	Withey *et al.*[10]	12	-
2001	Greuse *et al.*[12]	16	2
2003	Deunk *et al.*[4]	27	21
2004	Muzaffar *et al.*[13]	681	20
2006	Barboza *et al.*[14]	13	2
2010	Lumenta *et al.*[11]	26	42
2010	Cortez *et al.*[15]	72	5

Regarding gender distribution results show a predominance of cases in boys, frequent presence of bilateral and symmetric compromise, the third space being the most affected, which is in agreement with literature data. More than one third of the patients had some genetic syndrome, and in cases with no associated syndromes 61% of the cases syndactyly was accompanied by other malformations of the hand. These data show that, almost always, the treatment involves a multidisciplinary approach and with various surgical steps. Among all cases there is a slight predominance of simple syndactyly, mostly cases of easier surgical approach and better results.[16,17]
The set of data collected allows further studies to assess the correlation between age at surgery, technique used and malformations and syndromes associated with the aesthetic and functional quality of the final result.

CONCLUSION

The results obtained in this case series are very similar to those found in the literature, which validates the study and leads us to a better understanding of this condition in our midst.

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