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Epidemiological and Clinical Aspects of Cleft Hand: Case Series From a Tertiary Public Hospital in São Paulo, Brazil

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Abstract

Background: Cleft hand is a rare and congenital deformity that affects hands and feet and can be associated with other malformations. The objective was to evaluate the epidemiological and clinical aspects of cleft hand patient in a case series. **Methods:** Baseline characteristics associated with this deformity, such as sociodemographic characteristics, affected upper limb side, family history, clinical manifestations, and the degree of deficiency according to Barsky, Manske and Halikis, and Valenti classifications, were analyzed in 38 patients treated in the Department of Orthopedic Surgery of the Irmandade da Santa Casa de Misericórdia de São Paulo, Brazil. **Results:** A predominance of typical hands as classified by Barsky, types II and IV by Manske and Halikis, and type IV by Valenti was found. A high frequency of typical cases (55.3%), as defined by Barsky, had a positive family history ($P = .031$) and were associated with other clinical manifestations (44.7%), when compared with atypical cleft hand patients ($P < .001$). **Conclusion:** In our study, there were more typical cleft hands than atypical, and they were more commonly associated with family history and other clinical manifestations.

Keywords: congenital malformations, central ray deficiency, cleft hand

Introduction

Ectrodactyly, cleft hand, split hand, and lobster claw hand are synonyms to describe congenital central ray deficit of the hands.[13] The "V" shape of the cleft characterizes this anomaly that may be associated with the absence of one or more digits.[7,18] This anomaly is a longitudinal failure of formation considering the classification of congenital limb formations[16] and a I. Malformations, B. Abnormal axis formation/differentiation—hand plate, 4. Unspecified axis, iii Complex, c) Cleft hand of the Oberg, Manske, and Tonkin (OMT) Classification.[17]

Because of its broad spectrum of manifestations and rarity, treating cleft hands is challenging. This condition is genetically heterogeneous, with a sporadic etiology, or it may be associated with several clinical manifestations, such as EEC syndrome (ectrodactyly, ectodermal dysplasia, and cleft palate), congenital heart disease, and eye deformities.[11,12]

Since Barsky's[1] pivotal article, dated from 1964, many authors[1,4-6,8,16] have created different classifications based on cleft foot information, family history, and the association with other clinical manifestations.

The aim of this study is to report both epidemiologic and clinical data from a case series of 38 patients with ectrodactyly treated at the Division of Hand Surgery and Reconstructive Microsurgery of the Department of Orthopedic Surgery at the Irmandade da Santa Casa de Misericórdia de São Paulo, Brazil.

Materials and Methods

Study Design and Sample

A total of 38 patients (50 malformed hands) treated at the Hand Surgery and Reconstructive Microsurgery Division of the Orthopedic Surgery Department at Irmandade da Santa Casa de Misericórdia de São Paulo from 1990 to 2012 had their medical reports and images retrospectively gathered in a designated database for analysis.

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Table 1. Classification of Cleft Hand by Barsky.

	Typical	Atypical
Heredity	Common family history	No family history
Clinical presentation	Deep "V" shape cleft	"U" shape cleft more severe
	Occasionally with syndactyly	
	Bilateral	Unilateral
Cleft feet	Yes	No

Table 2. Classification of Cleft Hand by Manske and Halikis.

Type	Description
I	Normal web
II	Narrowed web
III	Syndactylized web
IV	Merged web
V	Absent web

Variables

Variables such as age, sex, unilaterality *versus* bilaterality of malformations, laterality of malformations (ie, right and/or left hand), as well as their classifications according to Barsky[1] (Table 1), Manske and Halikis[7] (Table 2), and Valenti et al[18] (Table 3) were appraised. Other clinically associated deformities, in particular cleft feet, cleft lips, cleft palate, EEC syndrome, Roberts syndrome, other teratogenesis, and cleft hand family history, were also logged into the database.

Barsky's Classification[1]

Barsky[1] considered typical cleft hands the ones with a deep "V" shaped deformity, occasionally associated with syndactyly and more commonly associated with cleft foot, family history, and bilateral involvement of members. Atypical cleft hands are characterized as "U" shaped deformity, are mostly unilateral and without involvement of the feet, and have no other clinical manifestations or previous family history (Table 1).

Manske and Halikis's Classification[7]

This classification is based, primarily, on the first web space. When the thumb web is normal, a type I is diagnosed; if the web is narrowed, a type II is present (mildly narrowed in *subtype a* and severely narrowed in *subtype b*); type III shows a syndactylized first web space, whereas type IV has a merged one; and, finally, type V shows no thumb web at all. The degree of the alteration to the thumb web space is directly related to the severity of the deformities found in the cleft (Table 2).

Table 3. Classification of Cleft hand by Valenti et al (2008).[18]

Group	Description	Subgroup
0	All fingers present, soft tissue cleft	
I	Normal web, third finger absent	a) Third metacarpal present b) Third metacarpal absent
II	Narrowed web, third finger absent	a) Third metacarpal present b) Third metacarpal absent
III	Syndactylized web	a) Simple syndactyly b) Complex syndactyly
IV	Merged web	a) Stable metacarpophalangeal b) Unstable metacarpophalangeal
V	Absent web	

Classification by Valenti et al[18]

The first type, as described by Ogino,[10] is type 0, in which all fingers are present and there is a soft tissue cleft among them. Type I shows a normal first web and absence of the middle finger, and has two different subtypes based on the presence of the third metacarpal, called *subtype a* when a third metacarpal present or *subtype b* if not. Type II is marked by a narrowed thumb web as well as the absence of the middle finger, being divided in two subtypes in the same fashion as type II. In type III, there is a syndactylized first web space (*subtype a* has a simple syndactyly, whereas *subtype b* has a complex syndactyly). When the thumb web is merged with the cleft, a type IV is diagnosed, called *subtype a* if a stable metacarpophalangeal is found or *subtype b* if unstable, and finally, type V is defined by the complete absence of the thumb web (Table 3).

Statistical Analysis

Categorical variables were analyzed through either chi-square or Fisher exact test when applicable. Yates' correction for continuity was also calculated. For all analyses, P values $< .05$ were considered to be significant. Both the Statistical Package for Social Sciences version 17.0 (SPSS Inc, IBM, Armonk, New York) and the Minitab version 16 (Minitab

Table 4. Presence of Cleft Hand Family History Among Typical and Atypical Deformities.

Cleft hand family history	Atypical	Typical	*P* value
No (%)	17 (100)	17 (80.95)	.031
Yes (%)	0 (0)	4 (19.05)	

Table 5. Presence of Other Clinical Manifestations Among Typical and Atypical Deformities.

Other clinical manifestations	Atypical	Typical	*P* value
No (%)	15 (88.2)	7 (33.3)	<.001
Yes (%)	2 (11.8)	14 (66.7)	

Inc, State College, Pennsylvania) were used to perform the statistical analysis.

Results

Among the 38 patients studied, over half of whom were men (21 patients, 55.3%), there was an identical number of patients presenting bilateral deformation in those exhibiting left malformation (12 patients, 31.6%, in each group), whereas a larger group had only their right malformed (14 patients, 36.8%). At the end, a total of 50 hands had been diagnosed with cleft hand. Only 4 patients (10.5%) presented family history of ectrodactyly, whereas the vast majority had none (34 patients, 89.5%). Other clinical manifestations such as split foot, cleft lip, cleft palate, teratogenesis, EEC syndrome, and Roberts syndrome were observed in 16 individuals (42.1%).

In regard to Barsky's classification, 21 (55.3%) patients were classified as having typical cleft hands, whereas 17 (44.7%) had atypical malformations (Table 1). Of the latter, only 1 had bilateral involvement and 2 presented other clinical manifestations. We found significant statistical differences between patients with typical and atypical deformities, concerning the presence of other clinical manifestations, $P = .031$ (Table 4), and family history of cleft hands, $P < .001$ (Table 5). All 4 patients with central ray deficit family history had typical cleft hands.

Of the 17 female patients, 10 (58.82%) had atypical hands; meanwhile, of the 21 male patients, 14 (66.7%) had typical cleft hands. Such difference was found to be of statistical significant difference ($P = .01$) (Table 6).

Because the classifications by Manske and Halikis[7] and by Valenti et al[18] are used only for typical cleft hands, the following data included only the description of 32 hands from 21 patients.

Seven out of 32 hands (21.9%) were classified as type I of Manske and Halikis,[7] 9 of 32 (28.1%) as type II (2 of them type IIa and 7 type IIb), 7 of 32 (21.9%) as type III, and 9 of 32 (28.1%) as type IV. There were no malformations considered as type V of the Manske and Halikis classification.

Table 6. Sex Distribution in Patients With Typical and Atypical Deformities.

Barsky	Typical	Atypical	*P* value
Male (%)	14 (66.7)	7 (33.3)	.01
Female (%)	7 (41.18)	10 (58.82)	

Similarly, no type 0 or type V of the Valenti et al[18] classification was observed in the study population. There were, however, 5 type Ia, 2 type Ib, 4 type IIa, 5 type IIb, 3 IIIa, 4 IIIb, 8 Iva, and 1 IVb hands according to this classification.

A higher frequency of positive family history for cleft hands ($P = .031$) and the presence of other concomitant clinical manifestations ($P < .001$) were revealed in patients with typical cleft hands rather than in patients with atypical ectrodactyly according to Barsky's classification.[1]

Discussion

No consensus was ever achieved concerning the most commonly affected side or bilateral versus unilateral involvement.[1,3,4,7-9,18] Overall, 12 of the 38 study patients presented bilateral involvement; however, if one takes into consideration only the patients with typical cleft hands, as done in the majority of prior reports,[3,4,7-9,18] one would find that 11 of 21 patients were bilaterally affected, which would thus be a more frequent impairment than unilateral ectrodactyly.

Compared to Barsky,[1] which has portrayed a frequency of typical cleft hands as high as 52.6% of cases, this study shows a predominant incidence of such cases, more specifically in 55.3% of patients from the studied case series. The occurrence of associated malformations in this analysis (42.1%) was rather similar to that described by the previously mentioned author in 1964, which was 47.4%.[1]

The results acquired from our investigation also support the preceding reports[2,3,14] which emphasized that atypical cleft hands are actually a variety of symbrachydactyly, as we were able to establish that such kind of malformation is remarkably less coupled with feet involvement, the presence of other concomitant clinical malformations ($P < .001$), and positive family history for cleft hands ($P = .031$).

Interestingly, in the present study, there was a statistically significant difference between sex distribution and Barsky's[1] typical and atypical deformity patients, which has not yet been reported.[1,4,7,18] One has to ponder whether such novel finding might be an effect of the number of patients assessed in this study.

To suit our data for comparison with more recent findings, atypical cleft hand patients were excluded from the analysis. Consequently, 14 patients (66.7%) from a population of 21

typical ectrodactyly patients had accompanying malformations. Adding to the controversy found in the literature, some authors fail to mention the actual number of associated malformations.[12] Falliner,[4] like others, considers cleft foot to be a part of the "cleft hand syndrome" and only credits, as concomitant associated defects, other limb and nonlimb malformations. Had cleft foot not been pondered as an associated malformation in the present study, there would only be 5 out of 21 (23.8%) patients presenting with additional concomitant clinical manifestations, a rate similar to that described by Manske and Halikis[7] (25%).

Split foot in patients with cleft hand was present in 9 of 21 analyzed patients (42.9%). Distinctive frequencies have been reported, being occasionally higher, as shown by Falliner,[4] who described 70% of cleft feet in patients with bilateral involvement of the hands or occasionally lower, as stated by other authors.[5,15]

The distribution of cleft hands presently found contrasted with that described by Manske and Halikis,[7] in which type III was the most frequent (34.3%), as well as with that portrayed in Falliner's[4] study, which showed type V as the most common variation. Both types II and IV were responsible for 28.1% of cases each in our case series, and no type V patients were present in it, which was also the rarest type in Manske and Halikis's[7] study and is exceptionally difficult to treat.[6] It is possible that the index fingers are always hypoplastic in type III patients, but measurements were deemed inappropriate in this study to achieve a definite conclusion.

The Valenti et al[18] publication showed type IV (compiled of both types IVa and IVb) as the most common, similar to our findings. Neither type 0, first described by Ogino,[10] nor type V patients were found in our cohort. Due to its rather complex features, and its single distinctive benefit of operating as a reminder of the importance of metacarpophalangeal stability in type IV patients, its daily usage remains somewhat impractical.

Strengths

Despite its nature of case series report, the present analysis remains as one of the largest samples of cleft hands in the literature to date.[1,4,7,8,18,14] More clinical information was provided on each case, unlike what is provided by the most common classifications.[1,4,7,18]

Limitations

As expected in retrospective studies, unlike what happens in prospective studies, there were no initial study design or a treatment algorithm based on either the classification or other aspects of cleft hands when the analyzed data first began to be put into a dedicated database. Moreover, because patients were treated in a wide time period of time (from 1990 to 2012), the long-term results regarding postoperative cosmetics, mobility, and strength might have been incomplete, and as techniques developed over the observed timeline, comparisons among them were also unattainable.

Conclusions

Typical Barsky cleft hands were more commonly associated with family history of cleft hands as was the presence of other concomitant clinical manifestations. Patients with either narrowed or merged thumb web and Manske and Halikis types II and IV, respectively, had a higher incidence in the present study.

Ethical Approval

This study was approved by our institutional review board.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Because we have only retrospectively evaluated medical reports and photos, there was no need for obtaining informed consent from the patients for being included in the study, according to institutional and national ethical committees.

Declaration of Conflicting Interests

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References

1. Barsky AJ. Cleft hand: classification, incidence, and treatment. *J Bone Joint Surg Am*. 1964;46(8):1707-1720.
2. Buck-Gramcko D. Cleft hands: classification and treatment. *Hand Clin*. 1985;1:467-473.
3. De Smet L. Classification for congenital anomalies of the hand: the IFSSH classification and the JSSH modification. *Genet Couns*. 2002;13:331-338.
4. Falliner AA. Analysis of anatomical variations in cleft hands. *J Hand Surg Am*. 2004;29:994-1001.
5. Hoyme HE, Jones KL, Nyhan WL, et al. Autosomal dominant ectrodactyly and absence of long bones of upper or lower limbs: further clinical delineation. *J Pediatr*. 1987;111:538-543.

6. Kay SP, Wiberg M. Toe to hand transfer in children. Part 1: technical aspects. *J Hand Surg Br*. 1996;21:723-734.
7. Manske PR, Halikis MN. Surgical classification of central deficiency according to the thumb web. *J Hand Surg Am*. 1995;20(4):487-497.
8. Nutt JN, Flatt AE. Congenital central hand deficit. *J Hand Surg Am*. 1981;6:48-60.
9. Ogino T. Cleft hand. *Hand Clin*. 1990;6:661-671.
10. Ogino T, Minami A, Kato H. Clinical features and roentgenograms of symbrachydactyly. *J Hand Surg Br*. 1989;14: 303-306.
11. Pries C, Mittleman D, Miller M. The EEC syndrome. *Am J Dis Child*. 1974;127:840-844.
12. Rasenmann A, Shapira T, Cohen MM. Ectrodactyly, ectodermal dysplasia and cleft palate (EEC syndrome). Report of a family and a review of literature. *Clin Genet*. 1976;9: 47-53.
13. Reed MH, Genez B, Chapter 19: Hands. In: Reed MH, ed. *Pediatric Skeletal Radiology*. Baltimore, MD: Williams & Wilkins; 1992:584-625.
14. Sandzen SC. Classification and functional management of congenital central defect of the hand. *Hand Clin*. 1985;1:483-498.
15. Sener RN, Sayli BS, Isikan UE, et al. Tetra-oligodactyly with bilateral aplasia and hypoplasia of long bones of upper and lower limbs: a variable manifestation of the syndrome of ectrodactyly with tibial aplasia. *Pediatr Radiol*. 1990;21:57-61.
16. Swanson AB. A classification for congenital limb malformations. *J Hand Surg Am*. 1976;1:8-22.
17. Tonkin MA, Oberg KC. The OMT classifications of congenital anomalies of the hand and upper limb. *Hand Surg*. 2015;20(3):336-342.
18. Valenti P, Lozano Gonzales E, Vergara E, et al. Cleft hand: a review of 33 cases and new ideas about classification. *Chir Main*. 2008;27:121-128.