

Original Article

A comprehensive functional classification of cleft hand: The DAST concept

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ABSTRACT

Context: Phenotypic expressions of the congenital cleft hand are variable and might baffle even the experienced as to the choice of surgery. The morphological parameters defining the anomaly dictate not only the functional capacity of the anomalous hand but also the degree of possible surgical restoration. Despite a large number of classifying systems available, none encompass all the relevant issues.

Aims: The purpose of this work is to present an all-inclusive and universally acceptable classification of the deformity which would graphically represent the entire gamut of possible presentations; principal and associated. Furthermore, based on such staging, the choice of surgical procedure and the stages of surgical intervention can be standardised to ensure the best results for the patient.

Patients and Methods: This study is based on a series of 27 patients with a total of 38 cleft hands.

Results and Conclusions: The necessity to include and to assess all determinants of function and complexity in these hands to standardise the choice of management, gave rise to the DAST system of classification; an acronym for all the morphological determinants of the anomaly (D = Digits missing, A = Associated anomalies in the hand, S = Site of cleft, T = Functional state of the Thumb). Numerical values were assigned to each component in increasing order of complexity. Score for each determinant as well as the aggregate score indicates the degree of complexity in a graphic manner. The DAST classification has a predictive value in choice of procedure and prognosticating surgical outcomes.

KEY WORDS

Classification; cleft hand; diagnosis; surgical management

INTRODUCTION

Congenital cleft of the hand described earlier as 'Lobster' or 'Claw' hand is defined as a longitudinal central deficiency that expresses itself as suppression of the bone and/or associated soft tissues in

the central elements of the hand comprising of the index, middle and ring fingers.^[1,2]

Our understanding of the development and genetics of the condition has gradually evolved with the earlier

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concept of the atypical cleft hand now clearly included within the symbrachydactyly sequence.^[3-6] The typical cleft hand, however, is still posing problems in terms of an accurate and comprehensive description of the anomaly as a whole. The source of this difficulty can be attributed to the variability in the number of rays suppressed, the site of the cleft-central, radial or ulnar and, the possibility of associated anomalies such as polydactyly, syndactyly or even disorganization and/or fusion of osseous components. Moreover, the functional status of the thumb by its involvement in the cleft, associated syndactyly/polydactyly or hypoplasia/aplasia; all presented either singly or in combination, further complicates the picture.

The aim of this work is to present a simple, all-inclusive and universally acceptable classification of the deformity which would graphically represent the entire gamut of possible presentations; both principal and associated. Based on such staging, the choice of surgical procedure and the stages of surgical intervention can be standardised to ensure the best results for the patient.

Cleft hand classifications

Given the unpredictability of phenotypic presentations of this anomaly, it is hardly surprising that a large number of classification systems have been proposed variously based on; the number of defective rays,^[7-9] teratologic mechanism of aplasia and synostosis,^[10] status of the thumb web,^[11] presence of associated anomalies^[12] and radiological morphology and cleft position— central, radial or ulnar.^[13] The teratologic classification is more of geneticists' interest and not very useful to the surgeon. Currently, the most accepted clinical classification (without prejudice to the other morphologic based ones) is the one given by Manske and Halikis.^[11] This classification is founded on the status of the thumb web, assuming it to be the primary predictor for the functional and aesthetic outcome in the anomaly. However, it fails to consider other morphological parameters such as associated anomalies, the effect on function due to the absence of more than one digit and, site of the cleft. Clefts of the hand, unfortunately, are too complex and multivariate in their presentations for taking such a simplistic, unifocal approach. Other morphological based classifications also suffer similar shortcomings.

The prime indication for correcting a hand deformity has to be the restoration of prehensile function to as near normal and as early as is possible. The aesthetic correction may be a natural spin-off of the surgery. The surgeon, therefore,

needs a functional system of classification with multivariate analysis which would provide a graphical representation of the anomaly, guide to the necessity as well as the choice of surgery and be of help in prognosticating outcomes in any phenotypic variant.

PATIENTS AND METHODS

In a series of 27 patients with a total of 38 cleft hands seen between 2002 and 2015, 16 were with unilateral cleft hands and 11 with bilateral. Seven patients were females and 20 males with ages ranging from 3 months to 64 years. Familial inheritance was seen in only 3 cases. The presence of cleft feet was associated in 7 patients of which 6 had bilateral cleft hands. A brief description of the cases is presented in Table 1.

A few diverse presentations which do not find a place in the erstwhile classification systems, thus emphasising the need for a comprehensive system, are shown in Figures 1-10.

DAST; the comprehensive functional concept

To be clinically efficient, a classification of the anomaly should ideally encompass all the possible morphological variants, be numerically indicative of the severity of the disorder, unambiguously streamline the choice of surgical procedure(s) and finally, enable the surgeon to realistically predict the results.

Table 1: Brief description of cases

| Number of cases | Type of cleft | Comments |
|------------------------|--|---|
| 18 | Central cleft with a missing middle finger | |
| 6 | Two digits missing | |
| 7 | Three digits missing | One patient had a unilateral 'paddle hand' with the absence of all digits except the thumb and little finger and the presence of a syndactylous web between the border digits |
| 4 | Four digits missing | Three with thumb present (synpolydactyly in one) One with only the ulnar border digit present |
| 2 | Cleft hands with features of extensive effects of congenital constriction ring syndrome in multiple digits in addition to syndactyly | |
| 1 | Very wide central cleft due to the presence of a transverse metacarpal along with syndactyly of thumb and index finger | There was radial cleft in six hands of which one had bilateral cleft hand. Ulnar cleft was seen in two hands. One child with bilateral cleft hand showed a radial cleft in left hand with one digit missing and an ulnar cleft in the right hand with polydactyly |

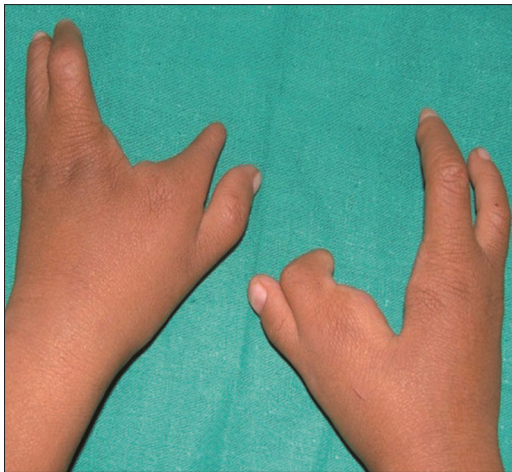


Figure 1: Central cleft with contracted 1st web (left side) and partial syndactyly of thumb and index finger (right side). Associated anomalies seen are Streeter's dysplasia (bilaterally for index fingers) and left-sided syndactyly of ring and little fingers. (D-1, A-5, S-1, T1 left/T2 right; aggregate-8/9) applicable surgical procedures- Type 2 (Snow and Littler/Oberlin procedure in view of wide cleft)



Figure 2: Central cleft with normal thumb and first web. Two suppressed rays and syndactyly between digits bordering the cleft. (D-2, A-2, S-1, T-0; aggregate-5); applicable surgical procedure-Type 2 (release of syndactyly followed by cleft closure)

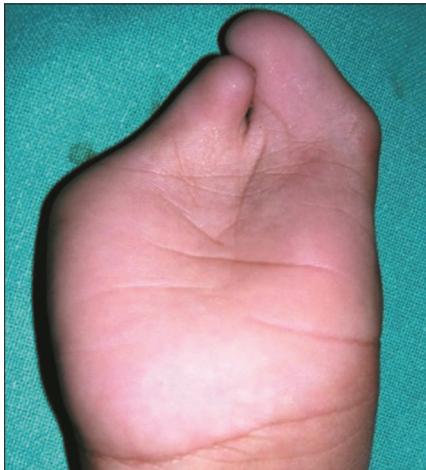


Figure 3: Central cleft with three missing rays with extensive syndactyly between thumb and little finger resulting in a 'Paddle Hand' (D-3, A-2, S-1, T-3; aggregate-9) applicable surgical procedures- Type 3 (release of syndactyly followed by toe transfer if warranted)



Figure 4: Complete synpolydactyly inclusive of the thumb associated with a central cleft of the hand (D-4, A-2, S-1, T-3; aggregate-10) applicable surgical procedure - Type 3 (toe transfer)

The morphological determinants of function for the cleft hand include the number of missing digits, associated anomalies of the hand, site of the cleft and, functional status of thumb. The authors have, over years of experience, evolved a comprehensive numerical system of classification in order of increasing complexity called the DAST system; an acronym for the morphologic determinants of the anomaly as follows:

Digits (number missing) 'D' - The number of metacarpals in hand affects the span of the hand. Surgical closure of the cleft becomes more difficult and may not even be an option if more than one digit is suppressed. The missing digits can be numerically graded from D1 to

D4 [Figures 1-4]. Clefts can be present without suppression of a digit^[14] or even with polydactyly^[15] [Figures 5 and 6]; both designated as D0.

Associated anomalies 'A' - These complicate the presentation and make management difficult either by necessitating staged or ancillary surgeries or, by adversely affecting the functional outcome. They can be numerically graded from 0 to 5 in an increasing order of complexity for surgical procedures and an attendant reduction in the likelihood of satisfactory functional and aesthetic outcomes [Figures 1-4 and 6-8].

The site of the cleft 'S' - These can be central, radial or ulnar. The surgical procedures for each type of cleft, based on its site, are different. Central is the



Figure 5: Ulnar cleft of the hand with no suppressed rays (D-0, A-0, S-3, T-0; aggregate-3); surgical management applicable - Type 1(translocation radially of ulnar fingers)



Figure 6: Ulnar cleft hand associated with polydactyly (D-0, A-1, S-3, T-0; aggregate score-4). surgical procedure applicable-Type 1 (translocation radially of ulnar fingers with or without addressing polydactyly)



Figure 7: Radial cleft of the hand with associated camptodactyly affecting two digits (D-1, A-3, S-2, T-0; aggregate-6) surgical effort may be directed to address the camptodactyly. No need to address the cleft



Figure 8: Central cleft of the hand with multiple associated anomalies (syndactyly 2nd and 3rd digits with abnormal fusion of bones) and a Pouce flottant thumb (D-0, A-5, S-1, T-5; aggregate-11) applicable surgical procedure- Type 3 (toe transfer for thumb reconstruction)

most common and easiest to close surgically with the best surgical and aesthetic outcomes expected; hence scored as S1 [Figure 9]. Radial clefts are the next in frequency. Standing alone they may not worsen the function, but when associated with other anomalies they make the management more complicated and are therefore scored as S2 [Figure 10]. Ulnar clefts are more complex, though rarer, and may require osteotomy of the carpus for radial transfer of the ulnar fingers to close the cleft,^[16] and are therefore scored as S3 [Figures 5 and 6].

Functional state of the Thumb 'T' - Thumb controls almost 50% of the functional capacity of the hand. The importance of a normal thumb for functional restoration in hand cannot be overstated. Although the classification by Manske and Halikis^[11] of cleft

hand is completely based on the state of the thumb web, a prudent view would be to grade this web from T0 (representing a functionally normal thumb) to T5 (indicating an absent or functionally useless thumb) [Figures 1, 3, 4, 8, and 9].

Thus, in the DAST system, numerical values have been assigned under each head in an increasing order of complexity [Tables 2-5]. The collective expression of these values describes the anomaly accurately, much like the TNM system for tumours.

All the morphological aspects of the anomaly are covered to enable the surgeon to decipher at a glance the complexity of the anomaly and the kind of surgical procedure(s) required.

Illustrative examples show the logic and simplicity of this system [Figures 1-10].

Table 2: DAST; numerical scale for number of missing digits 'D'

| Numerical score | Number of digits missing in the hand |
|-----------------|--|
| 0 | None |
| 1 | One missing digit |
| 2 | Two missing digits |
| 3 | Three missing digits |
| 4 | Four digits missing with only a border digit remaining |

Table 3: DAST; numerical scale for associated anomalies 'A'

| Numerical score | Presence of associated anomalies |
|-----------------|--|
| 0 | None |
| 1 | Polydactyly |
| 2 | Syndactyly |
| 3 | Constriction ring syndrome/contractures/abnormal bone fusion |
| 4 | Wide cleft with transverse bones in the cleft |
| 5 | More than one associated anomaly present |

Table 4: DAST; numerical scale for site of cleft 'S'

| Numerical score | Site of cleft |
|-----------------|---------------|
| 1 | Central |
| 2 | Radial |
| 3 | Ulnar |

Table 5: DAST; numerical scale for the functional state of the thumb 'T'

| Numerical score | Functional status of the thumb |
|-----------------|--|
| 0 | Normal, with no functionally significant anomaly |
| 1 | Contracted web without syndactyly |
| 2 | Partial syndactyly |
| 3 | Complete syndactyly |
| 4 | Hypoplastic thumb (Blauth I-III) |
| 5 | 'Pouce flottant' or thumb agenesis (Blauth IV-V) |



Figure 9: Wide central cleft of hand with transverse bone in the cleft and a contracted thumb web without syndactyly (D-1, A-4, S-1, T-1; aggregate-7) applicable surgical procedure - Type 2 (excision of transverse bone with Snow and Littler/Oberlin procedure for wide cleft)

By happy coincidence, 'Dast' in Persian refers to the 'hand' and makes the acronym even more appropriate and appealing!

Our experience over years of using such a classification permits us to recommend that as a thumb rule, an overall score >4 or individual score in any determinant >2 indicates an increasingly complex deformity with possibly less than satisfying functional and aesthetic long-term post-operative results.

DISCUSSION

The observation: 'Functional triumph but social disaster' by Platt^[17] considering cleft hands, continues to be misconstrued as an endorsement for limiting the role of surgery for aesthetic considerations. While function is undoubtedly the most important attribute needed in hand and the primary indication for surgery, it is desirable (though not always achievable) that the aesthetic deformity should not draw undue attention and cause social embarrassment.

Functional goals of surgical correction would be the restoration of a pinch, effective grasp and a good grip. Morphological presentation due to the interplay of various determinants is actually the greatest predictor of functional restoration in hand. It should, therefore, be the rationale for classifying cleft hand deformities provided such anomalies are considered comprehensively and not by determinants in isolation. Further, given the tremendous variability of presentation, no single surgical procedure can be applicable universally for all variants. The procedures primarily meant for the central cleft cannot apply for the ulnar one. The radial cleft would



Figure 10: Radial cleft of the hand (D-1, A-0, S-2, T-0; aggregate-3) no surgical management indicated

rarely require an intervention unless there is extensive syndactyly between the thumb and the digit at the ulnar border of the cleft. Other associated anomalies may require separate procedures. It is therefore imperative to understand the cleft comprehensively before making an appropriate surgical choice.

Depending on the status and complexity of the cleft as per the DAST classification, the hitherto available surgical procedures can be classified into Types 1-3:

Type 1 - Procedures applicable for D (0, 1), A*, S (1, 3) and T 0 clefts.

The aim is to close the cleft and create a physiologic web. The recommended procedures include:

- Creating a commissure; Barsky^[18]
- Translocation radially of ulnar fingers (TRUF).^[16]

The TRUF procedure is appropriate for the ulnar cleft (S3) but can be sometimes used for the wide central cleft (S1) with a prerequisite of a normal thumb, index finger and a normal web.

The number of missing digits has to be from none to one (D0-D1) and associated anomalies may or may not be present (A*). When present, they have to be tackled simultaneously or separately depending on their complexity.

Type 2 - Procedures applicable for D (0, 1, 2), A*, S 1 and T (1, 2, 3) clefts.

The aim here is to close the cleft, release the adducted thumb and create a physiologic thumb web.

They are indicated in patients with suppressed rays ranging from none to two (D0 to D2).

Associated anomalies may or may not be present (A*). The thumb ranges from a contracted web to almost complete syndactyly (T1 to T3 or Manske Type IIa to Type III). These procedures address the S1 (central) clefts and are not applicable for ulnar clefts. The web contracture or syndactyly may cause rotation of the thumb and may need an additional de-rotation osteotomy of the thumb metacarpal.

The eponymous procedures available can be listed in order of progressively deteriorating functional status

of the thumb: Miura and Komada,^[19] Ueba,^[20] Snow and Littler^[21] and Oberlin *et al.*^[22]

Type 3 - Procedures applicable for all clefts with the status of D (>2), A (1–5), S (1 or 2 with extensive syndactyly between thumb and ulnar digit) and T (>3) clefts.

The aim is the restoration of as effective pinch and grasp as possible.

These clefts have too many rays suppressed. A hypoplastic, ‘Pouce flottant’ or absent thumb and associated anomalies ranging from the simplest problem of polydactyly to multiple anomalies may also be present. The deformity is too severe to surgically attempt or hope to achieve a hand functionally or aesthetically anywhere near normal. The associated anomalies, as in the previous types of procedures, have to be tackled by an adjunctive procedure, carried out simultaneously or in a staged manner. A S2 (radial) cleft with suppression of the index and extensive syndactyly between the thumb and the next ray will require the release of syndactyly and creation of the thumb web by using locoregional flaps.

The question of whether or not to address the anomaly is paramount here since a good outcome is not assured. A considered decision needs to be made based on the principle of ‘primum non nocere.’ That said, the D4 as well as the T4, T5 hands require a toe transfer to at least provide a pincer grip by providing two border digits.^[23] However, this may not always be possible for the want of appropriate motors in these hands or the presence of associated cleft feet. The removal of transversely lying bones within the cleft is essential even if the hand has only the border digits since the divergence between these rays will be aggravated with growth. It is relatively easier to convince a parent for surgery if the deformity is getting worse, whether due to extensive syndactyly or the presence of transverse bones. Simultaneously addressing the cleft may be ideal.

It needs to be understood, however, that the decisions on the choice of procedures and whether to operate or not will be individual surgeon's choice based on all the above inputs regarding the anomaly and the expectations of the outcome in individual patients. The choice is not easy by any means. The above classification of various operative procedures available is only an attempt to bring clarity, by associating with the extent of the severity, in the clutter of eponymous procedures for the anomaly

with no relationship to the variant of the anomaly. None of these have, or can have, universal application in this anomaly of myriad presentations.

CONCLUSIONS

The DAST classifying proposal has universal applicability providing a comprehensive functional overview of the anomaly. It is easy to understand and communicate, creates a visual impression and permits easy recording both in documents as well as in digital form. The numerical grading, both of individual determinants and the aggregate score, graphically describes the anomaly and helps reaching a decision about the necessity, types and timing of surgeries. It has an excellent prognostic value which helps to communicate expected outcomes. The DAST system can seamlessly form a basis for standardising surgical management and comparing follow up results at the same or different centres engaged in the management of cleft hand.

Acknowledgements

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Conflicts of interest

There are no conflicts of interest.

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