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Guest Editorial

Addressing the neglected landscape of congenital hand anomalies

J. Terrence Jose Jerome, FRCS(G), EDHS[1], Khalid I. Khoshhal, MD.[2]

[1]Department of Orthopaedics, Hand and Reconstructive Microsurgery, Olympia Hospital and Research Centre, Trichy, Tamil Nadu, India, [2]Department of Orthopedics Division, Surgery, Prince Mohammed Bin Abdulaziz Hospital, Ministry of National Guard-Health Affairs, Almadinah Almunawwarah, Saudi Arabia.

***Corresponding author:**
J. Terrence Jose Jerome, FRCS(G), EDHS
Department of Orthopaedics, Hand and Reconstructive Microsurgery, Olympia Hospital and Research Centre, Trichy, Tamil Nadu, India.

terrencejose@gmail.com

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NEGLECT AND UNDERREPORTING

Despite advancements in medical science, the field of congenital hand anomalies remains largely untraversed.[1-3] This neglect stems from underreporting, lack of established treatment protocols, and absence of comprehensive guidelines and strategies. The variation in congenital hand anomalies across different geographical regions further complicates diagnosis and management.[2] More concerning is the frequent association of these anomalies with systemic conditions, such as heart and lung diseases, which often goes undiagnosed and inadequately addressed.

CASE EXAMPLE

A poignant example is a recent case involving a 21-year-old female with a congenital absence of both flexor and extensor tendons in her middle finger.[1] This rare condition involved the missing flexor digitorum profundus, flexor digitorum superficialis, and extensor tendons; yet, radiographic imaging revealed well-formed joints. The affected finger displayed hypoplasia and lacked both volar and dorsal skin creases, underscoring the complexity of congenital hand differences and the necessity for precise diagnosis and tailored management.

This case is particularly notable because the patient only sought medical attention due to the societal pressures surrounding marriage proposals. This highlights a common scenario where individuals with congenital anomalies seek corrective surgery before significant life events. The absence of treatment protocols means that decisions must be made on a case-by-case basis, based on the surgeon's experience, often without the benefit of comprehensive research or established guidelines. In this case, options such as ray amputation or staged tendon reconstruction were considered. However, the complexities of the patient's impending marriage influenced the decision to avoid surgical intervention, emphasizing the need for realistic expectations and minimizing surgical risks.

LACK OF DOCUMENTATION

The lack of detailed documentation and research on congenital hand anomalies significantly hampers progress in this field. Previous studies have extensively documented congenital bilateral

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absence of flexor digitorum superficialis in the little finger and flexor pollicis longus in the thumb. However, cases such as the total absence of both flexor and extensor tendons in the middle finger remain underreported.[1] Such omissions in the literature prevent the development of effective treatment strategies and hinder our understanding of these anomalies' broader implications.

Adding to the challenge is the scarcity of surgeons specializing in these anomalies worldwide. Congenital hand and upper limb anomalies are more prevalent in developing countries, where the high cost of treatment often prevents patients from seeking specialized care. This financial barrier contributes to underreporting, as individuals may either forgo treatment altogether or seek help from less specialized surgeons who are less likely to report these cases. Many congenital hand anomalies do not fit into the universal classification systems currently described, making diagnosis and treatment even more complex. What hand surgeons encounter in practice is merely the tip of the iceberg, and the cases they operate on are even rarer.

Furthermore, there are congenital anomalies in babies that do not survive long enough to be described and reported. The absence of a systematic reporting mechanism for these cases leaves a significant gap in our knowledge. Pediatricians play a crucial role in diagnosing these anomalies and ensuring early referral to hand surgeons, which is essential for timely and effective management.

STATISTICS AND STUDIES

A study in the United States identified 4,883,072 live births between 1992 and 2010 in New York State, including 2,314,661 births in New York City and 2,568,411 in New York State.[4] Among these, 13,278 upper extremity anomalies were identified, resulting in a prevalence rate of 27.2/10,000 live births, or 0.272% of newborns (95% confidence interval 0.267–0.277%). The study detailed specific anomalies: Polydactyly (21.6/10,000), Syndactyly (0.94/10,000), Apert syndrome (0.068/10,000), and several others with varying prevalence rates. Similarly, the reported incidence of congenital upper limb anomalies (CULA) in population-based studies from Australia and Sweden is around 20/10,000.[5]

ESTHETIC AND SOCIAL PERCEPTIONS

Esthetic hand preferences often favor youthfulness, health, proportionality, and symmetry. Factors such as abnormal finger number or length, multiple abnormal fingers, and evident hand or wrist abnormalities can negatively impact social perceptions. Cultural differences also influence these perceptions. A study involving Finland, Austria, and Singapore[5] evaluated the social impact of congenital hand anomalies and found that hands with intact symmetry were perceived better than those with disrupted symmetry. The least esthetically pleasing hands were radial club hand, monodactylous symbrachydactyly, and transverse reduction at wrist level due to a combination of negative factors.

Children were more critical of esthetics than adults, particularly rating ulnar polydactyly lower. This difference may stem from adults focusing on pre-operative hand normality and fearing post-surgical functional impairment, rather than solely esthetic concerns. Interestingly, clinodactyly received a higher mean score from children, possibly due to dirt under the fingernails in the normal hand photo. There were some sex differences; females generally gave higher scores to all hands than males and rated six-finger hands better than syndactyly, opposite to male respondents. Cultural differences were also noted; Singaporeans rated hands lower than Europeans, possibly due to cultural preferences for bodily wholeness. Participants familiar with limb anomalies, personally or through someone they know, rated their hands more favorably. Despite age, sex, or nationality, perceptions of congenital hand malformations were similar. Opinions varied least for the most and least pleasing hands, with cultural preferences influencing specific ratings.

ADVANCES AND CHALLENGES

Less is known from other countries, highlighting the need for global research and reporting on social, cultural, esthetic, functional, and many more aspects of their life. However, there is good news. Over the past 50 years, the clinical management of CULA has evolved significantly.[5,6] New disorders have been described, and approaches to treatment and reconstruction have improved. Clinical genetics has become an integral part of the diagnostic and management approach, leading to the identification of underlying etiologies for many CULAs. With these advances and an increasing emphasis on international collaboration, there is a move toward using international standards for healthcare diagnoses, outcomes, communication, and discoveries.

The International Federation of Societies for Surgery of the Hand (IFSSH) has shifted to the Oberg, Manske, Tonkin (OMT) classification scheme from the original Swanson-IFSSH classification of CULAs. There is good progress in this area, and there are benefits to a more standardized and collaborative approach. Despite these advancements, many congenital anomalies do not fit into the new classification system, emphasizing the need for continuous reporting and publishing of new CULAs to develop comprehensive treatment guidelines.

COLLABORATIVE EFFORTS

To address the persistent issues, it is crucial to establish robust reporting mechanisms, develop standardized

treatment protocols, and formulate comprehensive guidelines for managing congenital hand anomalies. Collaborative research efforts across geographical regions are essential to understand better the variation in these anomalies and their associated systemic conditions. By documenting and sharing rare cases like the one described, we can expand the spectrum of known congenital hand disorders and improve patient outcomes through informed clinical considerations.

Congenital hand anomalies represent a critical yet neglected area of medical science.[5,7] Enhanced reporting, standardized treatment protocols, and comprehensive guidelines are imperative to advance our understanding and management of these conditions. Only through concerted efforts can we hope to provide better care for individuals affected by these rare and complex anomalies.[7-10]

In summary, while change can be challenging, it is also essential. Advances in our understanding of limb development and genetics have necessitated a re-examination of how CULAs are classified and reported. The OMT classification was developed to enhance global interdisciplinary communication, improve diagnostic accuracy, and foster ongoing research into the etiology of limb anomalies. Recent discoveries using the OMT-IFSSH classification have prompted suggestions for modifications to reflect better clinical experience, limb development, and genetic relationships. As our knowledge expands, updates to the classification will occur, allowing us to develop more precise subclassifications and ultimately provide better patient care.

CALL TO ACTION

Editors of hand surgery and related subspecialty journals should prioritize publishing more on CULA, as even individual case reports can greatly enhance our understanding of these complex disorders. All editors must report on CULA due to the insufficient current understanding of these rare and intricate issues. Comprehensive reporting is crucial to advance knowledge and improve patient care.

AUTHORS' CONTRIBUTIONS

JTJJ and KIK conceived and designed the study, conducted research, collected data, and wrote the initial and final draft of the article. All authors have critically reviewed and approved the final draft and are responsible for the manuscript's content and similarity index.

USE OF ARTIFICIAL INTELLIGENCE (AI)-ASSISTED TECHNOLOGY FOR MANUSCRIPT PREPARATION

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

CONFLICTS OF INTEREST

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