The findings of the current study showed:

The current study results revealed that there were Polydactyly is the most common validated by a thorough examination of medical records and available radiographs. Results

Background: In this research present an overview covering hand malformations in an isolated form or as part of a syndrome, based on anatomic and genetic observations and taking into account the significance of recent molecular and developmental data. Accordingly, the current research will focus on conditions with known molecular and genetic causes, refraining from a surgical classification. Objective: The current study was conducted to assess the Impact of Congenital Hand Anomalies on Consanguinity at Selected Surgical Units, King Abdul-Aziz University Hospital, Saudi Arabia Method. A Descriptive research design was utilized with implementation of retrospective research method. The aim of this study was to find out the most common congenital hand anomaly in King Abdul-Aziz University Hospital Jeddah and the association of family history and consanguinity with congenital hand anomaly.

Data collection was performed with utilization a retrospective epidemiological study in 70 cases all data in this study were validated by a thorough examination of medical records and available radiographs. Results: The findings of the current study showed that the about half of the patients presented to study setting were Saudis (55.1%), Yemenis comes in second place. Males represent 60.9% of the sample accounting for 42 patients. The majority of the patients were born in Jeddah. It was found that Polydactyly is the most common congenital hand anomaly seen in the clinic among the other anomalies accounting for 41 out of 69 patients (78.3%). Not all patients were reachable to obtain the missing information or some were undocumented, so we summarized them with different total numbers for each variable. Moreover, the data revealed that, (68.6%) were presented in the first 2 years of life, that give advantages include potential for growth, development, and better scarring. Regarding consanguinity and family history, it was positive in (60%) & (48%) respectively. Conclusions: The current study results revealed that there were Polydactyly is the most common congenital hand anomaly seen in the clinic among the other anomalies within the study setting.

Keywords: Congenital hand anomalies - Limb development - Malformation - Syndactyly - Polydactyly - Brachydactyly - Genetics-Duplication- Finger abnormalities - Forearm Abnormalities- Hand Deformity- Thumb abnormalities- Toe transfer

1. Introduction

Parental consanguinity, as a recognized risk factor for congenital anomalies, has mainly been studied with a focus on the types of parental relationships and their effects on genetic syndromes or birth defects in general. Congenital fibrosis of the extra-ocular muscles (CFEOM) is a heterogeneous group of disorders that may be associated with other anomalies. The association of a CFEOM syndrome with ulnar hand abnormalities (CFEOM/U) has not been reported to date. (Alpay, Nesibe & Buyukdogan, 2015).

According to Ariela, Michel, Shneor, Yutao & (2015), congenital limb malformations show evidence of a wide continuum of phenotypic manifestations and may take place as an isolated malformation and as part of a syndrome. They are individually rare, but due to their overall frequency and severity they are of clinical significance. Currently, increasing knowledge of the molecular basis of embryonic development has significantly improved our understanding of congenital limb malformations. Additionally, genetic studies have concealed the molecular basis of an increasing number of conditions with primary or secondary limb involvement.

Moreover, congenital limb malformations exhibit a wide spectrum of manifestations, phenotypic variability and genetic heterogeneity. For a review on skeletal malformations. They are individually rare, but due to their overall frequency and severity they are of clinical relevance. Congenital limb malformations may be caused by genetic or environmental factors disturbing the regular developmental program during embryogenesis. (Dharma Bhatta & Anwarul, 2014).

Pegah, Maryam & Bita (2014), the study of the molecular mechanisms underlying human genetic diseases, the analysis
of animal model organisms and the use of transgenic mouse technology have considerably helped elucidate the genetic basis of limb development and malformation in the last years. These findings have strongly influenced the genetic classification of congenital limb malformations. As well, a number of non-genetic conditions exist that are mainly caused by external factors such as amniotic bands, vascular disruptions or teratogens.

Unpredictably, many upper extremity malformations cause little functional deficit. Children develop prehension with hands as they are, and they usually are not self-conscious of difference until they become socialized in school. In contrast, parents may be dismayed by the appearance of an anomalous hand and may hope that surgery can create a "normal" hand. The hand surgeon treating children with upper extremity anomalies must offer surgery to improve the child's function and cosmetics, when possible, and counsel parents about what is and is not possible with surgery. (Sabbagh, et al., 2014).

Orcun, et al., (2013), mentioned that, early surgery is defined as that performed within the first two years of life. Advantages include the full potential for growth, development, and patterns of use; improved scarring; early incorporation of the reconstructed part; and reduced psychological affect. Disadvantages of early surgery are increased technical difficulty and possible increased anesthetic risk. Most surgeons perform these operations when the patient is in the second year of life but no later than when the patient enters school.

Congenital anomalies are deformities that are present at birth. Any type of deformity in a newborn infant can become a challenge for the child as he or she grows. Hand deformities can be particularly disabling as the child learns to interact with the environment through the use of his or her hands. The degree of deformity varies from a minor deformity, such as a digital disproportion, to a severe deformity, such as total absence of a bone. Early on consultation with a hand surgeon is an important part of the treatment process for the child born with a hand deformity. Even if reconstructive surgery is not a possibility, there are many different types of prosthetic devices that can be used to increase function. (Kandasamy, et al., 2012).

The classifications for congenital hand anomalies can vary. The classifications listed below have been accepted by the American Society for Surgery of the Hand. Classifications may change as more knowledge is obtained regarding each of the conditions. There are seven groups of deformities of the hand that will be discussed, including Problems in development of the parts, Radial club-hand, Ulnar club-hand and Failure of parts of the hand to separate, which include; Simple syndactyly & Complex syndactyly. (Bittles & Black , 2010)

Fowzan , (2010), mentioned that the aim of surgery for a congenital hand anomaly is to improve both function and appearance. Apart from the face, the hand is the only other part of the body on regular display. Independent living is largely dependent on good bimanual hand function. For example a large proportion of activities of daily living such as washing, dressing, and feeding consist of bimanual tasks. It is only when we temporarily lose the function of one hand that the significance of this becomes apparent. However, children with congenital hand anomalies adapt very well to limitations of hand function and can often find "trick" maneuvers to achieve essential tasks.

Anand & Alan (2008), stated that, as there is a wide variation in the types and severities of hand anomalies these cases are largely managed in specialized clinics. It is in this setting that the child and family will have access to long-term multidisciplinary care which includes input from geneticists, psychologists, therapists and children’s hand surgeons. For some children with more complex anomalies, psychological support can be as valuable as surgery to aid integration with and acceptance by their peers.

Congenital hand anomalies can occur as an isolated anomaly or as part of a syndrome. They can also be sporadic or hereditary. They are the commonest birth defects in infants after congenital heart diseases. They form a wide spectrum of disorders involving all tissue elements of the upper limb and can vary in severity from mild hypo-plasia to complete agenesis of the limb. The deformity is minor in most cases but about 10 % of cases require treatment. The commonest classification system used to describe them is the one proposed by Swanson in 1976. This has been modified and adopted by the International Federation for Surgery of the Hand. However there are several other classification systems described and recently the Japanese Society for Surgery of the Hand has modified the IFSSH classification. (Yunis, Rafig & Mumtaz, 2010).

2. Participants and Methods

A Descriptive research design was utilized with implementation of retrospective research method. The aim of this study was to find out the most common congenital hand anomaly in King Abdul-Aziz University Hospital Jeddah and the association of family history and consanguinity with congenital hand anomaly . The data collection was performed with utilization a retrospective epidemiological study in 70 cases all data in this study were validated by a thorough examination of medical records and available radiographs.

A total of 69 congenital hand anomaly cases were undergone for correction between January 1, 2005 and December 31, 2015 in one center King Abdul-Aziz University Hospital in Jeddah, Saudi Arabia. Patients who had isolated congenital anomalies of the foot and who had hand anomaly due to trauma or burn were excluded. Ethical approval was taken from Medicolegal and Ethical Committee at KAUH. Patients files were extracted using the diagnoses codes that involve congenital hand anomaly (Q68.1 to Q87.4), some of the files were electronic and the old ones were hard files, few data were undocumented in the hard files, so the authors tried to contact those patients family to review and complete the missed data, 24 patient’s families responded to phone calls, upon to that missed data was completed and diagnoses was confirmed for them, rest of the patient’s families changed
their phone number, so it was difficult to reach them. Patients’ data were collected in a data sheet during Jan and Feb 2016, the data sheet included two categories, demographical data and clinical data.

Moreover the data analysis was done by Chi Square test using “Statistical Package for Social Sciences” (SPSS program version 21). Qualitative variables were presented as frequency and percentage, and cross tables for quantitative variables. P value < 0.05 was considered a significant difference. Essentially, most of the cases found are polydactyly and syndactyly, they were analyzed according to gender, nationality, age at presentation, consanguinity, family history of CHA, uni/bilateral, right or left hand, associated feet anomaly, syndactylized fingers, radial or ulnar polydactyly and associated syndromes. Patients’ data were revised and confirmed from patients’ medical records, x-rays and operation reports.

3. Results

Table 1: Relationship between Consanguinity and nationality, type of congenital hand anomaly, affected hand, feet involvement and associated syndrome

<table>
<thead>
<tr>
<th>Consanguinity</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Positive</td>
</tr>
<tr>
<td>Nationality</td>
<td></td>
</tr>
<tr>
<td>Saudi</td>
<td>16</td>
</tr>
<tr>
<td>Non Saudi</td>
<td>11</td>
</tr>
<tr>
<td>Congenital Hand Anomaly Polydactyly</td>
<td>20</td>
</tr>
<tr>
<td>Syndactyly</td>
<td>5</td>
</tr>
<tr>
<td>Poly syndactyly</td>
<td>1</td>
</tr>
<tr>
<td>Macro syndactyly</td>
<td>1</td>
</tr>
<tr>
<td>Olig syndactyly</td>
<td>0</td>
</tr>
<tr>
<td>Affect hand</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Bilateral</td>
<td>15</td>
</tr>
<tr>
<td>Feet involvement</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>No</td>
</tr>
<tr>
<td>Associated Syndrome</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>No</td>
</tr>
</tbody>
</table>

Table 2: Relationship between family history of congenital hand anomaly and consanguinity

<table>
<thead>
<tr>
<th>Consanguinity</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Positive</td>
</tr>
<tr>
<td>Family History</td>
<td></td>
</tr>
<tr>
<td>Positive</td>
<td>17</td>
</tr>
<tr>
<td>Negative</td>
<td>5</td>
</tr>
</tbody>
</table>

*Statistical significant difference

The results of this study revealed that about half of the patients presented to King Abdul-Aziz University Hospital were Saudis (55.1%), Yemenis comes in second place. Males represent 60.9% of the sample accounting for 42 patients. The majority of the patients were born in Jeddah. It was found that Polydactyly is the most common congenital hand anomaly seen in the clinic among the other anomalies accounting for 54 out of 69 patients (78.3 %). As for the rest of the data illustrated in [Table 1] not all patients were reachable to obtain the missing information or some were undocumented, so we summarized them with different total numbers for each variable. Of the 51 patients with known age at presentation for surgery, 68.6% were presented in the first 2 years of life, that give advantages include potential for growth, development, and better scarring. Regarding consanguinity and family history, it was positive in 60% and 48% respectively. About half of patients had both hands involved (51.6%) and the majority of them had no associated anomaly (87.9%) and 62.7% without feet involvement. As regards polydactyly, 70.6% were ulnar polydactyly and 81.8% of the hands with syndactyly were simple.

With reference to consanguineous marriage and its correlation to congenital hand anomalies, 45 patients out of 70 were with identified consanguinity. The results suggest that consanguinity is more common among the Saudi population represented by 59.3% compared to 40.7% in non Saudis. Regarding affected hands and feet involvement among patients with consanguineous marriage, those who had bilateral hand affected with no feet involvement represent 55.6% (15/27). However, there was no significant different obtained on comparing consanguinity with the different variables showed in [Table 3]. In the matter of patients who have a family history of congenital hand anomaly and its relation to having a child with a subsequent anomaly, the data showed no significance in relating the different variables. Although there was higher percent (62.5) of patients with positive family history among Saudis (15/24) compared to non Saudis. 83.3% of patients with polydactyly are positive for family history (20/39). The 24 positive family history patients, 1 (4.2%) patient was known to have a specific syndrome. However, indicates that patients who have a family history of congenital anomaly of the hands are significantly higher among those with positive consanguinity (P < 0.05).

4. Discussion

The findings of the current research study revealed that In this study regarding sex, affected side and associated anomaly the results show that there’s was a slightly higher percentage of male 60% than females 40%. Only 10% (n=7) of children had associated syndrome of congenital hand anomaly, and 34.3% (n = 24) children had a family history of congenital hand anomalies. About age of presentation of the congenital hand anomaly patients, Most anomalies presented in the first year of life (44.2%), unfortunately the rest of patients were undocumented. Consanguinity, Polydactyly is more commonly seen in the clinic among the other congenital hand anomaly accounting for 54 out of 70 patients (77.1 %). The largest number of Congenital Hand Anomaly fall in polydactyly class, there are 34 patients with polydactyly out of 45.It was found that the current research results is congruent with the data carried out by Altunhan, Ali Annagür, Murat Konak, Sabahattin Ertuğrul, Rahmi Örs & Hasan Koç ,(2012), who reported that congenital upper limb anomalies affect 0.1–0.2% of all newborns. They are often isolated phenomena but can be associated with other
congenital anomalies and may be the only external manifestation of a syndrome. Knowledge of the treatment options is imperative to ensure appropriate referral and counseling.

Furthermore, the current study finding is consistent with the data of the study carried out by Mónica Ritter, Rosa Liascovich, Jorge López-Cameló & Eduardo E. Castilla (2001), in which their study of parental consanguinity in specific types of congenital anomalies, the authors analyzed the association between parental consanguinity and congenital anomalies, split, when possible, into clinical subtypes, in an attempt to obtain some insight into their recognized etiological heterogeneity. The material consisted of 34,102 newborn infants, affected by one of 47 selected congenital anomaly types, ascertained by the Latin-American Collaborative Study of Congenital Malformations (ECLAMC) during the period from 1967 to 1997. The consanguinity rate for each congenital anomaly type was compared with that of the population under study (0.96%), and the potentially confounding effect of six selected variables was controlled through a conditional logistic regression analysis for those congenital anomalies significantly associated with consanguinity.

Moreover, there is a congruent between the present study results with the findings reported by Harlap et al., (2008), who concluded that pre-occurrence rates for the same congenital anomaly in siblings of consanguineous and non-consanguineous cases were compared. A significant association with parental consanguinity was observed for three congenital anomaly types: hydrocephalus, postaxial hand polydactyly, and bilateral cleft lip +/- cleft palate, while three additional anomalies, namely, cephalocele, microcephaly, and hand + foot postaxial polydactyly, showed a positive association, but statistical significance disappeared after adjustment for confounders, probably owing to sample size reduction. The association between consanguinity and Down syndrome was mainly due to the confounding effect of maternal age, while for hydrops fetalis and 2 to 3 toe syndactyly, the observed positive association could not be tested for confounders due to sample size reduction.

Regarding the current study results in relation to consanguinity, it was found that patients have a positive consanguineous marriage, accounting for 74.1% of syndactyly patients. 55.6% of the consanguineous patients got bilateral hands anomaly. In relation between the syndromic patients with CHA and consanguinity, 5 patients (18.5%) are positive for consanguinity. And for family history, 20 patients out of 39 with polydactyly are positive for family history with a rate of 83.3%, 3 patients out of 8 with syndactyly are positive for family history with a rate of 37.5%, and 1 patient only with poly syndactyly has a positive family history with a rate of 42.2%. There’s no unexpected finding in this study. This study results is consistent with the data of the study reported by Miura, Nakamura & Horii(1994) , in the study of the position of symbrachydactyly in the classification of congenital hand anomalies, the authors conclude that, concerning the clinical features of 53 cases of intercalated hypoplasia and 113 cases of distal aplasia are reviewed and compared with each other and with 129 cases of syndactyly. Tri-, di- and monophalangeal symbrachydactyly, and adactyly with nubbin digits are consecutive anomalies. Furthermore, the found that there was no difference between the intercalated and terminal types in the sex ratio, which was 36:17, male: female (male 68%) in the intercalated type and 75:38 (male 66%) in the terminal type. The male incidence in syndactyly was 71% (92:37), higher than in the intercalated or terminal types.

5. Acknowledgment

Appreciation is hereby extended to all the participants for the current research study.

6. Conflicts of Interest

None declared.

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