PATTERN OF CONGENITAL HAND ANOMALIES IN THE PAEDIATRIC SURGICAL AND ORTHOPAEDIC OUT-PATIENT CLINICS OF A TERTIARY CENTER IN SOUTH-SOUTH NIGERIA: A 5-YEAR PROSPECTIVE STUDY

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ABSTRACT

Background: *Congenital hand anomalies are a wide array of congenital anomalies of poorly understood etiologies mostly affecting the fingers. They constitute 10% of all congenital anomalies seen in children. Emotional worries and the attendant resorting to wrong treatment options could arise from poor parental counselling. This study aims to present the pattern of congenital hand anomalies in the University of Port Harcourt Teaching Hospital (UPTH) over a five-year period.*

Method: *Patients presenting with congenital limb anomalies to UPTH, from January 2015 to December 2019 were consecutively sampled. Associations between the anomaly and demographic characteristics were analyzed with Chi-square and Fisher's exact test as appropriate. $P < 0.05$ was termed statistically significant.*

Results: *Fifty-two hand anomalies in 27 patients were studied over the 5-year period. The total number of paediatric surgical consultations from both clinics within the same period was 562 giving a prevalence of 9.3%. Polydactyly was the most common congenital hand anomaly (23%, n=12) followed closely by syndactyly (19.2%, n=10). The association between the identified anomalies and the gender of patients was not statistically significant ($P = 0.07$).*

Conclusion: *Congenital limb anomalies are fairly common in the study population. Data available for this study may create the needed platform for proper discussions between parents and surgeons on expected treatment endpoints which may improve acceptance of available treatment options and reduce delays in presentation.*

Key Words: *Limb anomalies, congenital limbs, pediatric limb anomalies, polydactyly, syndactyly*

INTRODUCTION

Congenital hand anomalies are a wide array of congenital anomalies of poorly understood etiologies mostly affecting the fingers. They constitute 10% of all congenital anomalies seen in children. While most babies adapt well to minor anomalies, severe types may cause functional limitations like delayed or deficient development of motor skills, limitations in basic life care skills and possible emotional deficiencies.

Treatment is aimed at achieving independent and optimal use of the affected limb and vary from use of splints, physiotherapy, surgery and in most instances, a combination of options. Surgical treatment is exacting for severe cases, especially when such patients present in late childhood or at adolescent and may require inputs from several surgical disciplines.

Limbs with unsuccessful surgical treatment endpoints and complex cases may require amputation and prosthetic rehabilitation. In the developing world, the required surgical disciplines for the treatment of complex anomalies may still be poorly practiced in most centers[1] leaving ablative options as the most viable treatment option. Rehabilitation after surgical ablation is either unavailable or quite expensive. This calls for education of mothers on early identification of the anomalies to achieve early presentation, thorough evaluation of the patients' treatment expectations and open discussions on possible treatment outcomes.

Watson Stewart[2] in his work on principles of management of hand anomalies has stated certain factors that should be key priorities in planning treatment. These include: timing of surgery, functional aim of treatment, expected broad cosmetic appearance as well as appropriate counselling of parents[2].

The most popular classification for congenital anomalies of the upper limb in children is based on the work done by Swanson et al. in 1983[3] and modified by the Congenital Malformations Committee of the International Federation of Societies for Surgery of the Hand (IFSSH)[4].

I. Failure of formation of parts (arrest of development)

   A. Transverse arrest (common levels are upper third of forearm, wrist, metacarpal, phalangeal)

   B. Longitudinal arrest (including phocomelia, radial/ulnar club hands, typical cleft hand, atypical cleft hand otherwise referred to as part of the spectrum of symbrachydactyly)

II. Failure of differentiation of parts

A. Soft tissue involvement

B. Skeletal involvement

C. Congenital tumorous conditions (includes radio-ulnar synostosis, symphalangism (stiff proximal interphalangeal joints (PIPJs) with short phalanges), camptodactyly, arthrogryposis, syndactyly)

III. Duplication

IV. Overgrowth

V. Undergrowth (thumb hypoplasia, Madelung's deformity (abnormal distal radial growth)

VI. Congenital constriction band syndrome

VII. Generalized skeletal abnormalities.

Though Watson[2] has suggested some level of heterogeneity within subgroups in this classification model and clear differences in hereditary patterns within deformities in the same groups, this classification system still retains global acceptance.

With deep belief in traditional values and practices within the sub-region, several interpretations are usually given to these anomalies in children[5], leading to emotional worries and the attendant resort to wrong treatment options. An analysis of such anomalies therefore will provide the needed data for policy formulation with respect to public enlightenment campaigns and possible change in treatment-seeking behavior. This study therefore aims to present the pattern of congenital hand anomalies in a tertiary center in Port Harcourt, south-south Nigeria over a five-year period.

METHODOLOGY

Patients presenting to the paediatric surgical and orthopaedic clinics within the period of study (January 1st 2015- December 31st 2019) with congenital hand deformities were consecutively sampled. Patients who did not receive treatment at the study center were also sampled. Only patients' aged 0-17years of age with any congenital anomaly affecting the upper limbs were included in the study.

Diagnoses were by clinical reviews (done by consultant paediatric surgeons and orthopaedic surgeons) in line with standard definitive descriptions with relevant radiologic evaluations where applicable.

The study tool was a structured questionnaire. Relevant sections of the tool addressed subjects' bio-data, presentation parameters, and pattern of associated anomalies as well as syndromic involvement. Clinical descriptions of deformities were as stated by the investigators and other surgeon colleagues. Questionnaire was expert-validated.

Surgical treatment and outcomes were not within the scope of the study.

Data obtained were entered into IBM SPSS Software (Armonk, NY, USA; 2015; version 23). Descriptive analysis on the statistics with respect to socio-demographics, presentation pattern and malformation patterns was done. Associations between the anomaly and demographic characteristics were also analyzed with Chi-square and Fisher's exact test as appropriate. $P < 0.05$ was termed statistically significant.

RESULTS

Fifty-two hand anomalies in 27 patients studied over the 5-year period. Male: Female ratio was 1.7:1. The total number of paediatric surgical consultations from both clinics within the same period was 562 giving a prevalence of 9.3%.

Pattern of Hand Involvement

Pattern of hand involvement as shown in Table 1 revealed than most patients had bilateral involvement. The right hand involvement was marginally more compared to the left hand.

Table 1: Pattern of hand involvement

SN	HAND INVOLVEMENT	FREQUENCY	PERCENTAGE %
1	Right	13	25
2	Left	9	17
3	Bilateral	30	57
4	Total	52	100

Syndromic Involvement

Most patients (81.5%) with congenital hand anomalies had associated syndromic involvement. Anomalies occurring sporadically were fewer (18.5%).

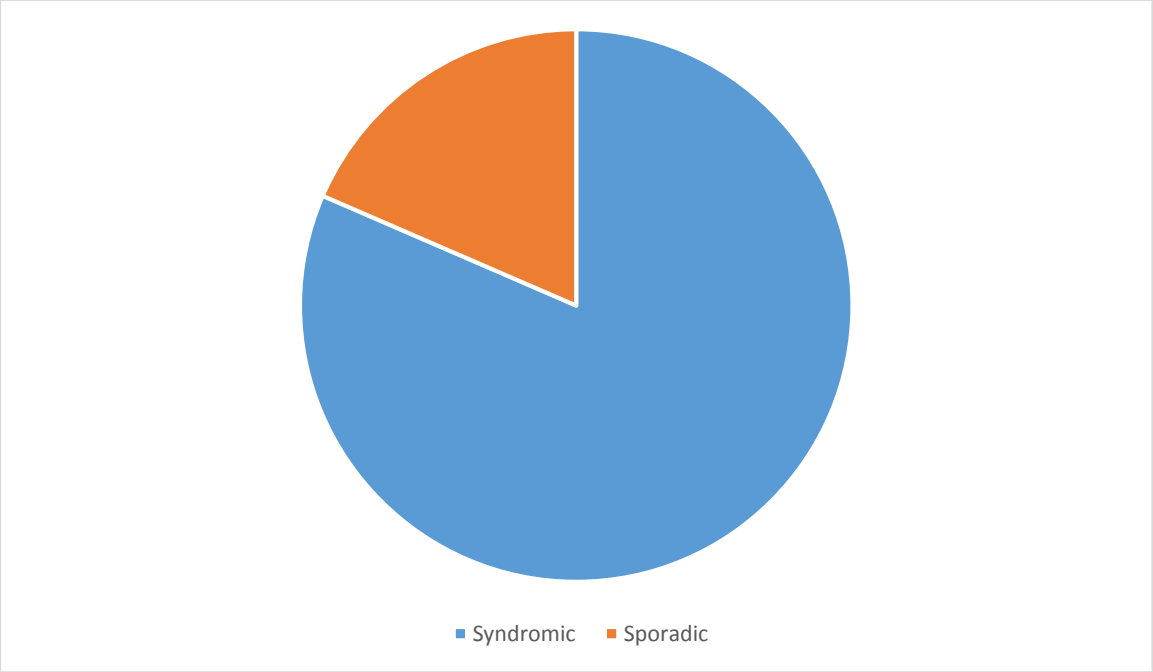

Figure 1: Syndromic Involvement

Types of Syndromes

The most common syndrome was Apert's syndrome accounting for 45.5% of all patients that presented with syndromic patterns. The other syndromes were Arthrogryposis multiplex congenita and Poland's disease.

TABLE 2: Types of syndromes

SN	SYNDROMES	FREQUENCY	PERCENTAGE %
1	Apert's	10	45.5
2	Arthrogryposis multiplex congenital	8	29.6
3	Poland	4	14.8
4	Total	22	100

Associated Anomalies

Umbilical hernias were the most common associated anomaly accounting 50% of all associated anomalies seen among the study subjects. Club foot was the most common lower limb anomaly associated with hand anomalies. Others are as shown in Figure 2.

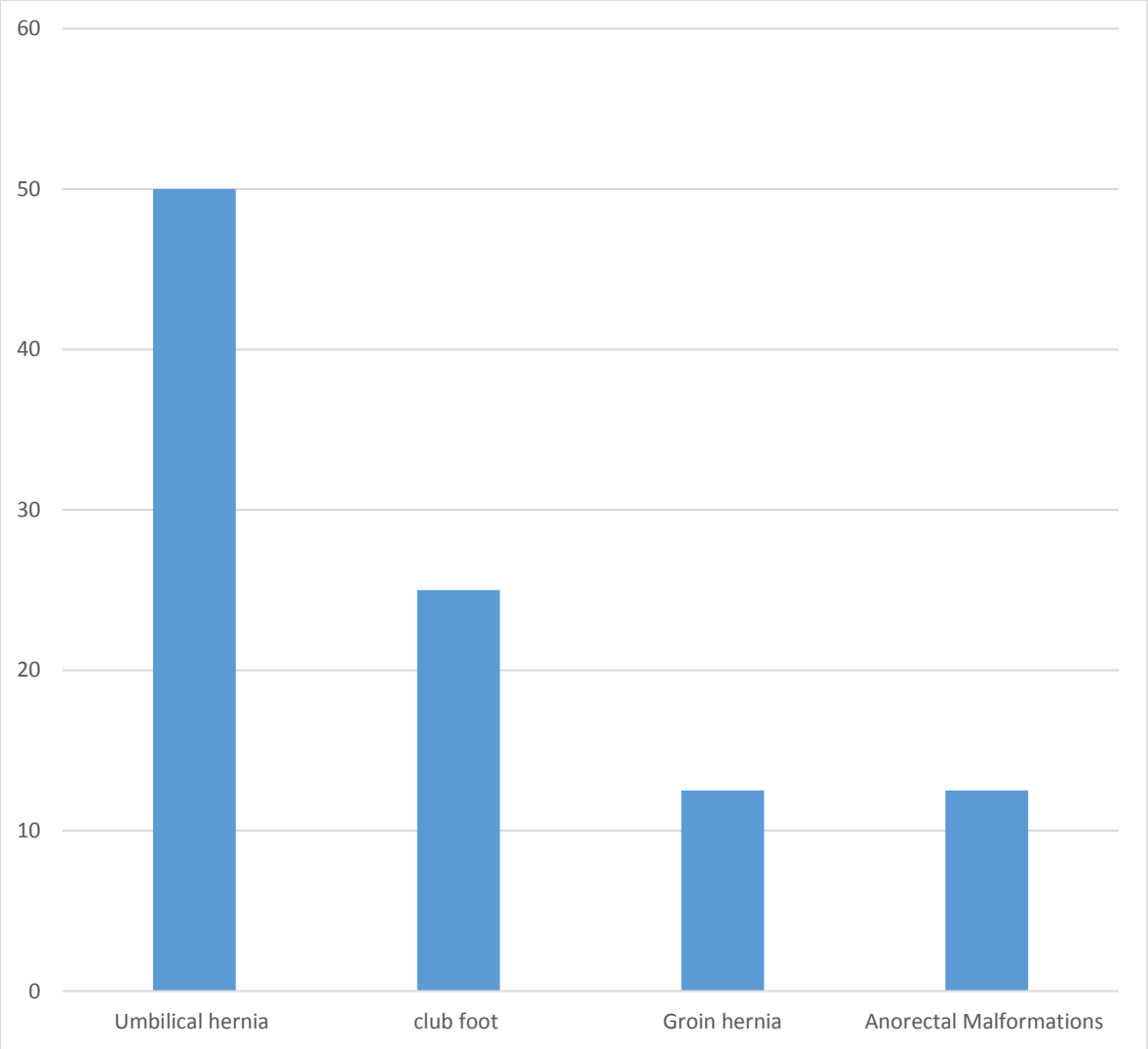

Figure 2: Associated Anomalies

Type of Congenital Hand Anomalies

23 percent of all congenital hand anomalies were type III anomalies (polydactyly). Syndactyly, constriction bands and brachydactyly were also common, accounting for 19.2%, 15.3% and 13.4% of all anomalies respectively. There were only 2 cases of cleft hand within the study period.

TABLE 3: Congenital hand anomalies

SN	HAND ANOMALIES	FREQUENCY	PERCENTAGE %
1	Polydactyly	12	23.0
2	Syndactyly	10	19.2
3	Constriction band	8	15.3
4	Brachydactyly	7	13.4
5	Campodactyly	5	9.6
6	Macrodactyly	4	7.6
7	Clinodactyly	4	7.6
8	Cleft hand	2	3.8
9	Total	52	100

The association between the identified anomalies and the gender of patients was not statistically significant ($P = 0.07$). There was also no statistically significant association between the type of anomaly and limb laterality ($P = 0.05$).

DISCUSSION

A total of 562 outpatients were seen in the paediatric outpatient and paediatric orthopaedic clinics within the study period. 52 congenital hand anomalies were seen in 27 patients within the same five-year period giving a prevalence of 9.3 %. Ajao &Adeoye[6] and Micheal et. al.[7] in similar studies in south-western Nigeria found a prevalence of 13.2% and 13.1% respectively. The former study was done among neonatal admissions which may probably account for the higher prevalence.

The slight male preponderance found was similar to findings by other workers within the region [5,6,7,8].

More than half of the patients (57%, n=30) had bilateral anomalies. This is similar to findings from the 10-year retrospective review by Micheal et al. which showed 54.9% bilaterality. Other authors[11,12,13] have also reported higher incidence of bilateral anomalies compared to unilateral anomalies. Though Giele et al[12] have reported more bilateral involvement in patients with generalized anomalies, Dao et al., have shown equal distribution of both unilateral and bilateral congenital hand anomalies.

Polydactyly was the most common anomaly (23%, n= 12) followed closely by syndactyly (19%, n=10) this contrast finding by Mba et al. in South-East Nigeria who found higher incidence of syndactyly than polydactyly. Their analysis was done in a regional referral hospital with plastic surgical interest. Most patients with polydactyly may have presented to the paediatric/orthopaedic surgical clinics. A five-year prospective study in South-East Nigeria with patient recruitment from several outpatient clinics from two tertiary centers reported polydactyl as the most common congenital anomaly[8]. A prospective study in southern Nigeria which recruited patients form the paediatric clinics, burns and plastic clinic as well as the postnatal clinics of a tertiary hospital also found polydactyly as the most common anomaly.

Only a quarter of patients had associated anomalies with the abdominal wall defects (umbilical hernias) being the most common. This contrasts findings by Michael et al.[6] in South West Nigeria who reported more foot anomalies. Their study was conducted in the plastic surgical unit of a tertiary center. Most patients with abdominal wall defects will be seen by the pediatric surgical clinic.

Most patients from this study had associated syndromes (81.5%, n=22). The huge percentage of syndromic patients may be a selection bias since most patients were recruited from the paediatric surgical outpatient clinic where syndromic patients with complex primary or associated deformities will usually present. Goldfarb et al.[9] in the United States reported only twenty percent prevalence of associated syndromes in a population-based study.

The country currently lacks birth defect registries. Their presence will help improve data collation and aid the estimation of a true prevalence in the population.

LIMITATIONS

The separation of the paediatric surgical sub-unit from the rest of the surgical department may influence the number of patients seen.

Patients that presented to the plastic surgical unit were not included in the study.

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Conflicts of Interest: None

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