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Original Article

Congenital upper limb anomaly as a cause of physical handicap



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ABSTRACT

Introduction: Congenital anomalies are most common causes of handicap in developing and developed countries. There are many approaches to classify congenital limb defects, especially upper limb.

Our study aimed to present a profile of the cases of handicap due to congenital anomalies of upper extremity encountered in B. S. Medical College & Hospital, Bankura, West Bengal. This may be helpful to the practicing orthopedic surgeons to assess and treat congenital anomalies of the upper extremity.

Methods: All the cases of congenital orthopedic anomalies affecting the upper limbs who attended the B. S. Medical College & Hospital in Bankura, West Bengal, for the purpose of obtaining physically handicapped certificate during a period of 1 year were included as subjects of the present study. All cases were subjected to clinical and radiological examination. Complete history was taken.

Results: Different types of deformities noted in our series, like transverse arrests at different levels, longitudinal arrests of preaxial variety, central longitudinal arrest (cleft hand) and intercalated longitudinal arrest or phocomelia. Overgrowth (macrodactyly), undergrowth (radial hypoplasia) both was noted. There was a case of constriction band syndrome.

Discussions: This study comprised of cases who attended hospital to obtain physical handicap certificate only and none had any therapeutic intervention. There was no case of total handicap. Incidence of congenital upper limb anomalies was higher in this series. More males were affected than females; there were more right sided defects than left. No definitive causes for the deformities could be isolated, chromosomal abnormality studies might have revealed etiology.

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1. Introduction

Congenital anomalies are abnormalities of structures of body parts arising at the time of conception, or during intra-uterine period. They are the most common causes of handicap in developing and developed countries.

Handicap is the disability which adversely affect normal growth, development & adjustment of life for a substantial period of life, if not permanently.

Congenital anomalies affect 1%–2% of new-borns, and approximately 10% of those children have upper extremity abnormalities. 1,2

Limb malformations can be categorized into 3 major groups:

1) A genetically-determined group, 2) An environmentally-induced group, 3) A multifactorial group.

There are many approaches to classify Congenital Limb Defects, especially upper limb. The most widely accepted classification of congenital limb anomalies was proposed by Frantz and O'Rahilly (1961)³ and presented by Swanson (1968).⁴ This work eliminated much of the confusing Greek and Latin terminology and has been accepted by the American Society for Surgery of the Hand (ASSH) and the International Federation of Societies for Surgery of the Hand (IFSSH) [1983], and the International Society for Prosthetics and Orthotics. This system defines the anomalies according to the embryonic failure during development and relies on the clinical diagnosis for categorization. Each limb malformation is classified according to the most predominant anomaly and is placed into one of seven categories, viz:

1.1. Type I - Failure of formation

- 1) Transverse arrest Can be at any level, shoulder to phalanx
- 2) Longitudinal arrest Preaxial Varying degrees of hypoplasia of the thumb or radius Central Divided into typical and atypical types of cleft hand Postaxial Varying degrees of ulnar hypoplasia to hypothenar hypoplasia
- 3) Intercalated longitudinal arrest Various types of phocomelia

1.2. Type II - Failure of differentiation

- Soft tissue Syndactyly, trigger thumb, poland syndrome, camptodactyly
- 2) Skeletal Various synostoses and carpal coalitions
- 3) Tumorous conditions Include all vascular and neurologic malformations

1.3. Type III - Duplication

May apply to whole limb, mirror hand, polydactyly.

1.4. Type IV - Overgrowth

Includes conditions such as hemihypertrophy and macrodactyly.

1.5. Type V - Undergrowth

Most commonly, radial hypoplasia, brachysyndactyly, or brachydactyly.

1.6. Type VI – Constriction band syndromes

Occurs with or without distal lymphedema; may involve amputation at any level.

1.7. Type VII — Generalized anomalies and syndromes

Ogino (1997)⁵ classified upper limb anomalies into

- i) Longitudinal deficiency radial, ulnar, central deficiency or cleft hand
- ii) Transverse deficiency
- iii) Constriction band syndrome

The clinician must possess a basic understanding of embryogenesis, limb formation, and inheritance patterns to relay relevant knowledge to the family. Certain upper extremity anomalies occur in isolation, whereas others are associated with systemic conditions. These associated disorders often take precedence over the limb anomaly and must be assessed with appropriate diagnostic testing.⁶

Our study aimed to present a profile of the cases of handicap due to congenital anomalies of upper extremity encountered in Bankura Sammilani Medical College & Hospital, Bankura, West Bengal. This may be helpful to the practicing orthopedic surgeons to assess and treat congenital anomalies of the upper extremity.

2. Methods

All the cases of congenital orthopedic anomalies affecting the upper limbs who attended the Bankura Sammilani Medical College & Hospital in Bankura, West Bengal, for the purpose of obtaining physically handicapped certificate during a period of 1 year (between August, 2008 and July, 2009) were included as subjects of the present study. The study population were residents of western parts of West Bengal and were either originally from western West Bengal or migrants from parts of surrounding states of Jharkhand and Odisha.

Clearance from Institutional Ethical Committee was taken and informed consent of the patients was taken before carrying out the study and taking photographs.

All cases were subjected to a thorough clinical and radiological examination. Complete history like maternal & paternal age, occupation, drug history, disease, injury, exposure to radiation, consanguinity, etc. and personal and family histories were taken. Chromosomal abnormalities were not searched for.

3. Results

In our study, we observed that 430 cases attended B.S.M. College to receive Handicapped certificate. Of them, 291 cases

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