Bilateral syndactyly of both extremities in a new born - a case report

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Abstract
Syndactyly is a condition where in two or more digits are fused together. A newborn female baby showed not only webbing of the fingers but also toes. X-ray of both hands and feet showed no bony fusion. Syndactyly is a rare anomaly that has surgical and psychological implications. Normally mesenchyme between prospective digits in hand and foot plates is removed by cell death (apoptosis) when this process fails the result is fusion between two or more digits. It occurs as one in every 2000 to 2500 births and twice as common in males. Though it is very rare it can be corrected surgically and child can live a normal life. The knowledge of digit formation and causes for abnormalities in the digits and toes are important for radiologists, obstetricians and surgeons for accurate diagnostic interpretation.

Keywords: syndactyly, zygodactyly, synpolydactyly, ringsmalldactyly.

Introduction
Syndactyly is a condition characterised by webbed digits, most often adjacent fingers. It occurs commonly in some mammals like kangaroo but is unusual condition in humans. The webbing of toes is normal pattern in many birds such as duck, amphianibian such as frog. Syndactyly can be partial or complete. In complex syndactyly the bones of the adjacent digits are fused. Isolated syndactyly is one of the most common congenital malformation of the hands and feet. On the anatomical basis, Tentamy and Mckusick have subdivided isolated syndactyly in to five types:.

Type 1 - Zygodactyly - between the middle and ring fingers (Most common form); Type 2 - Synpolydactyly - This also involves middle and ring fingers but includes a duplication of ring finger; Type 3 - Ring-small finger syndactyly - This is usually bilateral and occasionally the distal phalanges are fused. There may be only a rudimentary phalanx in the small finger; Type 4 - Hass-type syndactyly - In this rare type, there is complete syndactyly of all the digits. Occasionally a sixth metacarpal and phalanges may be included in the cup shaped hand; Type 5 - In this rare type both the middle and ring fingers, second and third toes are syndactylized. The fourth and fifth metacarpals and metatarsals may be fused.

The term syndactyly covers both webbed fingers and toes, 'webbed toes' also known as 'twin toes', 'duck toes' and 'tiger toes'. Syndactyly occurs when apoptosis or programmed cell death during gestation fails or is incomplete.

Syndactyly is associated most commonly with the following clinical conditions:

a) Syndactyly or Familial syndactyly b) Down syndrome c) It is also associated with a number of rare conditions notably Apert syndrome, Bardet-Biedl syndrome, Carpenter syndrome, Edwards syndrome, Miller syndrome and Timothy syndrome. A rare case of webbing of the fingers and also the toes in a new born is presented here.

Case Report
During a study concerning the incidence of congenital anomalies among the babies delivered at MNR Medical College and Hospital, Sangareddy for a period of six months starting from September 2011 to February 2012, a total of 2495 deliveries were observed. The total number of babies with congenital anomalies were 25, among this 11 were live born and 9 were still born and 5 fetuses were aborted. Among these, one female child was born alive with complete syndactyly, where both digits and toes were fused. Family history
of the mother or the father did not reveal any such case in their families. Genetic evaluation could not be done due to lack of availability of facilities. The investigations like x-ray and ultrasound were done.

Observations

The case reported here is a live born baby with complete syndactyly. On observation this case was found to have fused digits of both hand and foot.

On gross examination of the extremities, it was seen that (Fig. 1) all four fingers except thumb were fused together in both hands. All the toes in both feet were fused together. No associated anomalies were found in this case. X-ray of the hand and feet showed no bony fusion (Fig. 3 & 5).

On clinical examination it was seen that:

All four fingers except thumb were fused together in both hands, (Fig. 2) All the toes in both feet are fused together, (Fig. 4). This syndactyly is complete variety as fingers and toes are fused.

Discussion

Incidence and embryological basis: A limb anomaly is called dymelia, these include all forms of limb anomalies such as amelia, ectrodactyly, phocomelia, polydactyly, syndactyly, polysyndactyly, oligodactyly, brachydactyly. Luekin3 was the first person to describe the family which extends, over eight generations, comprising 338 individuals including 77
affected family members. In 23 individuals, among them syndactyly was present in both hands and feet, where as four individuals presented with malformation of only the feet. The present case also shows involvement of both hands and feet. Montagu6 has opined that occurrence of skipped generations indicate that penetration is < 100%. It is caused by failure, during sixth to eighth week of intrauterine life, of the usual longitudinal interdigital necrosis that normally separates the fingers. It happens either as an isolated anomaly like webbing of the digits or syndactyly. Sadler4 has commented that limb defects occurring in 6 per 1000 live births, with 3 - 4 per 1000, affecting upper limb. A different category of limb defects involves digits. If two or more fingers or toes fused it is called Syndactyly. Normally mesenchyme between prospective digits in hand and foot plate is removed by cell death (apoptosis). In one per 2000 births, this fails and the result is fusion between two or more digits4. At the fourth week of development, limb buds appear from ventrolateral body wall. The fore limb buds appear first followed by the hind limb 1-2 days later. In the six week old embryos the terminal portion of the limb bud becomes flattened to form hand and foot plates. Finger and toes are formed when cell death in the apical ectoderm ridge separates the ridge in to five parts. Further formation of digits depends on their continued outgrowth under the influence of the five segments of ridge ectoderm condensation of mesoderm to form cartilaginous digital rays and the death of intervening tissue between rays. Molecular regeneration of limb development is continued by HOX gene. Most of the malformations involving the limb abnormalities are due to embryonic insults between 4th and 6th weeks of gestation.

**Genetic basis of syndactyly:** Genetics play a major role in the formation of the digits, hence digital abnormalities occur if there is mutation.

**Five types of syndactyly and the corresponding loci associated with these types and their common phenotypical expression have been identified:**

Type I - 2q 34 - q36 - Webbing occurs between middle and ring fingers and / or second and third toe6.

Type II - 2q31- Also involves long and ring fingers but has a sixth finger merged in between6.

Type III- 6q21 - q23 - Small finger is merged in to the ring finger.

Type IV- 7q36 - involves all fingers and / or toes.

Type V - 2q31-q32 - Similar to type I, but metacarpals and metatarsals may also be fused.

The present case confirms to the Type 4 described above Castilla et al.,9 are of the opinion that syndactyly type I - also named "Zygodactyly" is inherited as an autosomal dominant trait and accounts for the majority of isolated syndactylies, with an incidence of 2-3/10000 in newborns. Riddle et al.,10 commented that Indian hedgehog (IHH) a homolog of Sonic hedgehog (SHH) which is an important morphogen in vertebrates plays a key role in establishing anteroposterior polarity of the limbs. Sarfarazi, Akarsu7 have described the localization of the syndactyly type II (synpolydactyly) locus to 2q31 region and identification of tight linkage to HOXD8.

Bosse, et all6 have described localization of Gene for Synacytly type I to chromosome 2q34-36, 2q31-q32, 6q21-q23 and 2q31 respectively where as Syndactyly type IV (SD4) is extremely rare and gene localization have not been assigned. The SD4 manifests complete syndactyly of all fingers with polydactyly and fusion fingers gives hand cup shaped appearance. A number of gene mutations have been identified that affects the limbs and some time other structures. The role of HOX gene in limb development is illustrated by two abnormal phenotypes produced by mutations in these genes. HOXA 13 results in hand foot - genital syndrome, characterized by fusion of carpal bones and small digits affecting females often have (didelphic) divided uterus. Mutations in XOXD 13 results in combination of syndactyly and polydactyly (synpolydactyly).
Surgical corrections of the syndactyly: Flat has described steps involved in surgical correction techniques employed for syndactyly. Syndactyly of the border digits (thumb/index finger or ring/small finger) is best treated at an early age to prevent longer digits curving towards smaller digit with the growth. Typically syndactyly is treated at six months of age. The treatment of syndactyly of the other digit is effective and more commonly preferred at 18-24 months of age, because the circumference of the conjoined fingers is smaller than the circumference of the two separated fingers, there is not enough skin to cover both digits once they are separated at the time of surgery. Vidyadharan and Lester have described the surgical procedure, aesthetic reconstruction of syndactyly correction.

Maternal causes: Man, Chang are of the opinion that the Maternal cigarette smoking during pregnancy increases the risk of having a child with congenital digital anomaly. The present study is a case of complete syndactyly, where all the fingers except thumb in upper limb and all the toes in lower limb are fused, which is a very rare entity, hence it is reported here. Present case coincides with the observations made by Sadler, incidence wise. While dealing with antenatal mothers, they must be educated about the intake of Folic acid, avoiding abuse of alcohol and cigarette smoking.

References:


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