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A RETROSPECTIVE COHORT STUDY IN MANAGEMENT OF STREETER'S DYSPLASIA: OUR EXPERIENCES

Plastic Surgery

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ABSTRACT

INTRODUCTION :-
Streeter's dysplasia is a sporadic condition that may also be present in association with other congenital anomalies. It has an incidence varying from 1 in 1500 to 1 in 15,000 live births. It is also known as congenital constriction band syndrome or amniotic band syndrome. This syndrome has significant predilection for the upper extremities and distal limbs.

AIMS AND OBJECTIVES:- The two main objectives of treatment of this study are
1) To improve function of the limbs
2) To improve cosmetic appearance

MATERIALS AND METHODS:-This retrospective cohort study was conducted at the Department of Plastic,Reconctructive and Faciomaxillary surgery at Madras Medical College and RGGGH between 2014 to 2018. 10 cases were included in this study ,out of which 4 males and 6 females. Age limit was 3 months-3years. The location and degree of each constriction ring and associated anomalies were recorded. 9 patients were treated with Z-plasty. In one patient due to tight deep band ,band excision with distal debulking was done.

RESULTS:- This retrospective cohort study conducted at the Department of Plastic,Reconctructive and Faciomaxillary surgery at Madras Medical College and RGGGH between 2014 to 2018 showed excellent improvement in the function of both upper limbs and lower limbs. Cosmetic appearance was good.The most common associated anomalies were syndactyly, acrosyndactyly, hypoplastic phalanges, lymphedema and Talipes Equino Varus.

CONCLUSION:-Most importantly, CCBS is not always fatal although there are many problems that come along with it. The future for those suffering from CCBS is continually improving reaching in utero surgical procedures. It is extremely important to seek prenatal medical attention when pregnant. This study concludes with ideal treatment option of Z-plasty for congenital band syndrome and improved limb function and aesthetic appearance of the patients with that procedure.

KEYWORDS

Congenital band syndrome, Streeter's dysplasia, Amniotic band syndrome

INTRODUCTION

Congenital constriction band syndrome(CCBS), also known as amniotic band syndrome, is a frequent cause of terminal congenital malformation of limbs, with a reported incidence of 1 in 5000 to 10,000 children. The clinical presentation is variable, with bands that may be superficial and incomplete or deep and circumferential, extending to the underlying bone. Multiple bands may be present in a single extremity. Distal to the site of constriction, vascular and lymph engorgement may lead to marked swelling and, in the case of deep, circumferential bands, significant neurovascular impairment.The distal aspect of the limb, particularly the longest digits, are most frequently affected. Amputations and syndactyly are seen in >50% of affected children.The incidence of clubfoot in constriction band syndrome ranges from 12%-56%. These feet often are rigid and more difficult to treat than idiopathic clubfoot, and typically require surgical treatment.

PATTERSON CLASSIFICATION:-

TYPES	FEATURES
TYPE1	SIMPLE RING CONSTRICTIONS
TYPE2	RING CONSTRICTIONS ACCOMPANIED BY DEFORMITY OF THE DISTAL PART WITH OR WITHOUT LYMPHEDEMA
TYPE 3	RING CONSTRICTIONS ACCOMPANIED BY FUSION OF DISTAL PARTS RANGING FROM MILD TO SEVERE ACROSYNDACTYLY
TYPE 4	INTRAUTERINE AMPUTATIONS

Surgery options for those suffering from CCBS include Z-plasty and W-plasty.When performing Z-plasty, "a z-shaped incision is made along the line of the greatest tension or contraction, and triangular flaps are raised on opposite sides of the two ends and then transposed.In contrast with Z-plasty, W-plasty the edges of the excised part are left in the form of a zigzag and the triangles are interdigitated for surgery .With both techniques there are great outcomes in relieving tension of band fixing both cosmetic appearance and function of the limb treated. In case of tight deep bands, band excision with debulking of distal segment can be done ,preserving limb adequate limb vascularity.Complications do exist as with any surgery including flap necrosis, wound infection and hematoma.

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PROCEDURE:-

Fibrous bands may be noted encircling the arms,fingers or toes of a child. They are simply removed or unwound, often untethering fingers from one another. Treatment of band indentation has consisted of band excision and soft tissue recontouring with multiple Z plasties . Release of circumferential bands was done to avoid vascular embarrassment to the distal segment. When bands are extremely tight, profound distal edema is managed by band excision and distal debulking was done in one case.Tight, deep bands may narrow the underlying bone. Skin and underlying fat may be separately transposed to avoid persistent indentation at the site of flap transposition. Deep fascial constriction is sought and released, particularly beneath deeper proximal bands. Proximal bands affecting the arm are relatively uncommon, but may be associated with distal neural or lymphatic compromise. The insensate hands that result from neural compromise are particularly vulnerable to infection and contracture. Those limbs with lymphatic inadequacy are vulnerable to repeated cellulitis.

CLINICAL PICTURES:

Case 1: CCBS Right Upper Limb

Figure 1: Preoperative Picture Of Ccbs Right Upper Limb

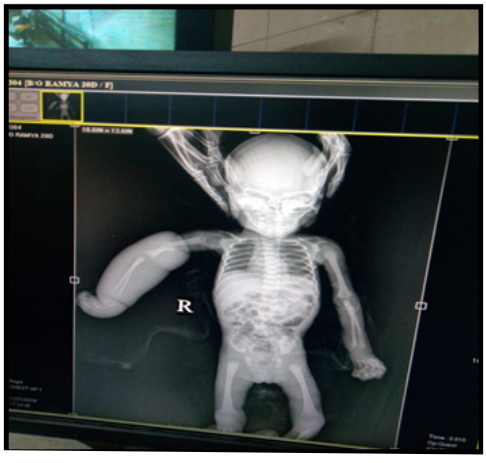


Figure 2: X-ray Image Of Ccbs Right Upper Limb

Figure 3: Z-plasty

Figure 4: Postop Image

Case 2: CCBS Lower Limb

Figure 5&6: Z-plasty In Lower Limb CCBS

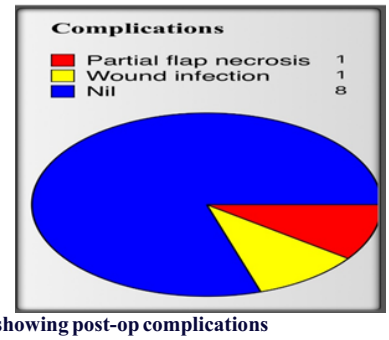
RESULTS:-

This retrospective cohort study conducted at the Department of Plastic, Reconctructive and Faciomaxillary surgery at Madras Medical College and RGGGH between 2014 to 2018 showed excellent improvement in the function of both upper limbs and lower limbs. Cosmetic appearance was good. In general the most commonly involved digits were middle and ring finger in upper limb. Most frequently involved toes were second and third toe. Intrauterine amputations were also common. The most common associated anomalies were syndactyly, acrosyndactyly, hypoplastic phalanges, lymphedema and Talipes Equino Varus.

Out of 10 cases , 2 cases developed complications like partial flap necrosis and wound infection but no disturbance in functional recovery at the end of one year of follow up period. All cases showed good functional recovery.

Table 1: Patient Details

S.NO	AGE	SEX	CCBS UPPER LIMB	CCBS LOWER LIMB	PROCEDURE	COMPLICATIONS	FUNCTIONAL RECOVERY	FOLLOW UP
1	6mon	M	YES	-	Z-PLASTY	-	GOOD	1 YEAR
2	1year	F	-	yes	Z-PLASTY	-	GOOD	1 YEAR
3	2years	F	yes	-	Z-PLASTY	-	GOOD	1 YEAR
4	5 mon	M	yes	-	Z-PLASTY	PARTIAL FLAP NECROSIS	GOOD	1 YEAR
5	1 year	F	yes	-	Z-PLASTY	-	GOOD	1 YEAR
6	8mon	M	yes	-	Z-PLASTY	-	GOOD	1 YEAR
7	6mon	F	-	yes	Z-PLASTY	WOUND INFECTION	GOOD	1 YEAR
8	1year	F	yes	-	Z-PLASTY	-	GOOD	1 YEAR
9	7mon	M	yes	-	Z-PLASTY	-	GOOD	1 YEAR
10	3years	F	yes	-	BAND EXCISION WITH DISTAL DEBULKING	-	GOOD	1 YEAR



Pie chart showing post-op complications

DISCUSSION

Congenital Constriction Band Syndrome (CCBS) is a controversial subject affecting fetus due to the fact that there is no known cause yet agreed upon. Many theories have been made regarding its cause and therefore CCBS may be referred to by other names. Some include amniotic band syndrome (ABS), also known as "Adam Complex", Streeter Dysplasia, annular groove, ring constriction syndrome and Pseudoainhulm[1].

This condition has been studied since the time of Hippocrates and Aristole in the early 300 BC. Some thoughts as to what could be the real cause of this condition include developmental problems in the formation of limb connective tissue, germ plasma defect, histology, and maternal trauma leading to a rupture of the amniotic membrane[2].

There is a significant predilection for the upper extremities and an increased frequency in distal limbs, and longer digits are significantly more involved than shorter ones[3]. "The hands are affected in almost 90% of cases [2]. Therefore, it is a condition that should be made aware of to educate pregnant women as well as individuals working in health care.

Constriction of appendages by amniotic bands can cause many troubles to the unborn fetus, and severity of the problem depends on where the constriction occurs and how tight it is. Some include constriction rings around legs, arms, and digits causing swelling of that body part, which inhibits blood flow. This leads to congenital lymph edema, or a blockage of fluid in a developing fetal lymphatic system. However, if the constriction is very extreme congenital amputation may occur as well. This decrease in blood supply may cause the effected body part to fall off completely, known as Amelia. Along with these effects come other abnormalities associated with CCBS, including clubfoot, clubhands, cleft lip, cleft palate, and hemangioma [4].

Theories :-

There are two main theories, **the first of which is the intrinsic theory proposed by George Streeter**, the director of embryology at Carnegie Institute, in 1930 which is called, "Streeter's Dysplasia." He believed a germ plasma defect was the cause of CCBS. His theory consisted of a disrupting event occurring during blastogenesis which leads to soft tissue sloughing off. Then, external healing of the slough begins leading to the constricting rings resulting in local developmental defects. He explained the ring constrictions as areas of defectively formed tissue due to defective germ plasma areas, and due to the closeness to the amnion caused its connection. This theory is supported by evidence of cases in which the infant affected is born with the amnion intact [1]. Also in support of this theory are the cases with renal and cardiac abnormalities that occurred in 37% of the cases in which fibrous bands from the amnion could not have reached [2].

Another theory, **the extrinsic theory was proposed by Richard Torpin,** an obstetrician, in 1965 and is known as the Amniotic Band Syndrome (ABS). His theory was first suggested by Hippocrates that maternal trauma lead to rupture of the amniotic membrane. Proceeding rupture, the amniotic sac stops growing properly and eventually separates from the chorion. The chorionic side of the amnion emanates numerous mesoblastic fibrous strings which entrap and catch the fetal parts. This then leads to the constriction of the body part stopping blood flow to that area leading to fetal amputations as one of many consequences . To better understand how this occurs, it is important to understand how a fetus develops in a mother's uterus. The fetus floats in the mother's uterus surrounded by amniotic fluid. The fluid is kept around the fetus and the uterus by a sac. This sac is composed by two layers adhered together. (See Fig. 7) .The outermost layer is the chorion and lines the uterus. The inner layer closest to the fetus is the amnion [5]. According to Michael Harrison (MD) from the Fetal Treatment Center 2009 this is the process in which ABS occurs: It is believed that amniotic band syndrome occurs when the inner membrane ruptures, or tears, without injury to the outer membrane. The developing fetus is still floating in fluid but is then exposed to the floating tissue from the ruptured amnion. This floating tissue can become entangled around the fetus [5].

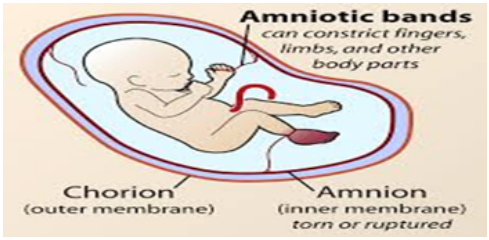


FIGURE 7:Image demonstrating fetus in amniotic sac consisting of two membranes.

Since the causes of CCBS are only theories, there is no known prevention. Some facts that have been studied show that CCBS have no sex predilection. It equally occurs to male and female fetuses. Nevertheless since many believe the cause of CCBS is amnion tearing which could accidently occur, other things could lead to this. For example, a "deformed uterus and a collagen disease of mother and fetus could predispose to this event" [6]. Although a collagen disorder may add to the risk of CCBS, it has not been proven to be hereditary. "The lack of family history or predictable recurrences in families of children born with ABS negates the theory of an inherent or genetic component to the condition. Amniocentesis or amniotic fluid test (AFT) is also thought to cause CCBS. AFT is, "a medical procedure used in prenatal diagnosis of chromosomal abnormalities and fetal infections" [7].

Zych, et al [8] in 1983 reported a case of involvement of congenital bands, pseudoarthrosis & impending gangrene of leg. They perfomed multiple Z-plasties reported the limb salvage. **Greene WB [9]** in his study advised a one-stage release for circumferential congenital constriction bands which was performed in four extremities (three patients).

A new technique, namely, **Mutaf procedure**, for the correction of congenital constriction rings.In this technique, following excision of the fibrotic constriction ring, the groove is filled with the turnover dermofat flaps elevated from both sides of the ring in an alternating pattern. Then skin closure is done with rectangular-plasty technique. A normal extremity contour will be obtained, with complete elimination of the sandglass deformity caused by the constriction ring.[10]

CONCLUSION:-

Most importantly, CCBS is not always fatal although there are many problems that come along with it. The future for those suffering from CCBS is continually improving reaching in utero surgical procedures. It is extremely important to seek prenatal medical attention when pregnant. This study concludes with ideal treatment option of Z-plasty for congenital band syndrome and improved limb function and aesthetic appearance of the patients with that procedure.

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