Congenital Constriction Band Syndrome and
It's Effects on Infants
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Abstract

The cause of Congenital Constriction Band Syndrome has been in debate over 300 years. There are two main theories as to what could be its true cause. The two main theories discussed are the intrinsic and extrinsic theories. This condition may cause inhibiting or life-threatening conditions as well as secondary deformities. To correct or help add function to these conditions there are surgical options available. Pregnant women should be informed of possible diagnosing options and treatments for the future of their infant.
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Congenital Constriction Band Syndrome (CCBS) is a controversial subject affecting fetuses 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 (Light & Ogden, 1993, p.143). 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 (Twee, 2009, p.1)

Whichever the cause, CCBS is estimated to occur in 1 out of 1,200 pregnancies to 1 in 15,000 live births in the U.S. It is also believed to be the cause of 178 in 10,000 miscarriages. It affects both sexes equally and is a congenital disorder that causes constriction of limbs, digits, or other body parts of a fetus. A congenital disorder describes a condition existing at birth or before birth (Twee, 2009, p.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” (Choulakian & Williams, 2008, p.221). “The hands are affected in almost 90% of cases (Twee, 2009, p.3). Therefore, it is a condition that should be made aware of to educate pregnant women as well as individuals working in health care.

Features
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 (Goncalves & Jeanty 1999).

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 (Light & Ogden, 1993, p.153). 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 (Twee, 2009, p.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 (Light & Ogden, 1993, p.153).

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. 1) The outermost layer is the chorion and lines the uterus. The inner layer closest to the fetus is the amnion (Harrison, 2009, p.2). 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 (Harrison, 2009, p.2)
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Today, this theory presented by Torpin is the most widely supported.

Problems

Some of the effects of these constrictions include indentations, amputations, acrosyndactylization, clubfeet, facial clefts, palate deficiencies, encephaloceles, thoracoschisis, along with many others could occur (Twee, 2009, p.1). The severity of the problem depends on the location and the tension of the band and if severe can be life-threatening for the fetus. For example a band becoming wrapped around areas such as the head or umbilical cord are areas of high concern (Harrison, 2009, p.3) Indentations occur when the fibrous bands were not tight enough to cause amputation of the limb. However, after the fetus is born this could lead to edema if not surgically corrected. (See Fig. 2) Amputations are prevalent when the limbs of the fetus were not able to grow due to the band entirely preventing blood flow to the area causing necrosis, or cell death. Additionally, in some cases these bands act as thread threading digits together resulting in a condition called acrosyndactylization. (See Fig. 3)
Acrosyndactylization can vary in severity from a few digits being bound together, to multi-digit. Constriction bands across the face and head may possibly lead to facial clefts, and if extended into the cranium encephaloceles can occur (Harrison, 2009, p.2). If these bands cross the body congenital fissure of the chest wall can happen, this is called thoracoschisis. (See Fig. 4) Although these conditions are of concern, the ultimate consequence of CCBS is amputation of some type or fetal death (Jones & Sinclair, 1988, p.169)

**Diagnosis**

Unfortunately, diagnosing of CCBS is most often difficult. Currently, ultrasound is the modality of choice due to its safe usage on fetuses. However, the fibrous strands are so small they are difficult to visualize. More often, 3D and 4D are being used to visualize suspected cases of CCBS due to its capacity to improve recognition and visualization of fetal anatomy and abnormalities. Magnetic Resonance Imaging (MRI) is also being used for its ability to view tissue structures in detail. Frequently, if bands are detected they are so indirectly due to swelling and constrictions of limbs (Harrison, 2009, p.3). “Neonatal diagnosis of ABS if frequently difficult and is accurately diagnosed in only 29% to 50% of cases” (Chandran, Lim, & Yu, 2000, p.12).

**Treatments**
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Treatment options for infants suffering CCBS usually occur after birth. Plastic and reconstructive surgeries are available to correct deformity and provide function of impaired part. Very rarely if CCBS is detected in utero, fetal surgery may be performed to save a limb or other deformity. However, this is usually not a consideration unless the fetuses’ umbilical cord or vital organs are in danger. This operation has been performed successfully on a 28 week pregnant woman. The earliest surgery of this type was done at Melbourne’s Medical Center in Australia, but has also been done successfully in the U.S. (Twee, 2009)

Surgery options for those infants 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, the edges of the excised part are left in the form of a zigzag and the triangles are interdigitated for surgery” (Choulakian & Williams, 2008, p.222). With both techniques there are great outcomes in relieving tension of band fixing both cosmetic appearance and function of the limb treated. Complications do exist as with any surgery including flap necrosis, wound infection, and hematoma.

In addition, other surgical options include the two-stage with direct closure approach. In direct closure the patient is under anesthesia and a tourniquet is applied to the limb. The limb is then

**Fig 5.** Arm (A) and foot (B) with deep constricting bands. C, D The same limbs following two-stage excision with direct closure

outlined with a surgical pen and must be meticulously dissected to avoid damaging the underlying neurovasculature. Then, the constriction band is excised and direct closure is done. Completion of closure allows the fatty tissue to naturally reposition itself under the skin. (See Fig. 5) With the two-stage approach, the first operation consists of one-half of the circumferences being excised and the other half being excised after three to six months. This method helps avoid problems to the distal circulation in the limb.

“Lymphedema, when present, will significantly improve within a few weeks of the first surgery” (Choulakian & Williams, 2008, p.223). Furthermore, in the case of syndactyly, staged release is used for improved cosmetic appearance and more importantly function. With multiple digit syndactylies, it may be possible to release all the distally joined finger tips in the first stage. This may require multiple stages for complete release. Full thickness skin grafting is harvested from the groin crease for complete coverage of distal finger tips. (See Fig 6) “Although staged release has been proved as safe and effective, it may be impossible to preserve all nerve quality” (Light & Ogden, 1993, p.147)

**Prevention**

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” (Jones & Sinclair, 1988, p.169). 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” (Twee, 2009, p.6).

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” (Young, Lindenbaum, Thompson & Pembrey, 1985, p.1063). This is done with Ultrasound guidance by a physician who inserts a needle through the abdominal wall of the mother and through the uterus until reaching the amniotic sac. Once there, they extract amniotic fluid to test the fetus’s cells for infections or abnormalities. This exam is usually performed at about the 18th week of pregnancy but may cause risk of damaging the infant’s limbs. Avoiding these exams and avoiding certain drugs such as cocaine and mifepristone may reduce risk of CCBS. All three are shown to increase chances of spontaneous rupture of amniotic membrane (Young et al., 1985, p.1063).

Conclusion

Although there are many unanswered questions regarding congenital constriction band syndrome, studies regarding its cause have came a long way. From 1652 J.B. van Helmont’s report of intrauterine amputations being attributed to pregnant mothers having looked upon maimed soldiers, to Torpin’s most supported theory of the amnion rupturing (Twee, 2009, p. 1). Whichever the cause, many surgical options are available in order to treat those suffering from CCBS. 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. Regardless what theory proves to be evident in causing CCBS, it is extremely important to seek prenatal medical attention when pregnant.
References


Young, I.D., Lindenbaum, R.H., Thompson, E.M., Pembrey, M.E. (1985) Amniotic bands in
connective tissue disorders. *Archives of Disease in Childhood, 60*(11) 1061-1063