A Case of Bilateral Craniofacial Cleft From Amniotic Band Syndrome

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Amniotic band syndrome [ABS] or amniotic disruption complex is defined as a destructive process of fetal damage in a random non symmetrical distribution, which is not attributable to aberrations in normal embryologic development processes .The mechanism has been considered to be initiated in rupture of the amnion, with the fetus then coming into contact with the chorionic surface of the amniotic membrane ,to which it adheres or becomes entwined by fibrous strands .As the fetus grows ,The tethered regions are progressively distorted, leading to constrictions, amputations ,Slash defects, and body wall or cranial defects .The severity varies from a single constrictive ring of a digit to lethal thoraco abdominal eviscerations or craniofacial distruction.ABS is a rare disorder in which bands of mesoderm that emanate from the chorionic side of the amnion and insert on the fetal body can lead to a wide variety of disfiguring and disabling malformations[1,2].

ABS results in the amputation of the fingers or limbs, associated with a wide spectrum of congenital anomalies usually involving the trunk and craniofacial regions including the skull. AbBS is in general sporadic and the incidence is approximately 1 in 15,000 live birth. Characterized by a destructive fetal process that is initiated by rupture of the amnion. Proposed as a sequel of intrauterine rupture of the amnion resulting in oligohydramnios and passage of fetus to chorionic cavity .The fetus subsequently becomes adherent to intertwined in, and tethered by fibrous mesodermic bands .As the fetus grows, its anatomy is distorted .This may lead to cranial or body wall defects or and although been reports of associated malformations of internal structures. [3, 4]

ABS that involves craniofacial region may present as cleft lip or palate , oblique or transverse facial cleft .These craniofacial anomalies in ABS are typically often bizarre and frequently non-embryological .The exact cause of ABS is still unknown .This lack of direct evidence has led to the proposal of two main etiological theories. The extrinsic and the intrinsic 4. The extrinsic theory states that the band of ruptured amnion causes an extrinsic compression which then results in constriction rings and other deformities of the developing fetus, while the intrinsic theory holds that germ cell deficiencies result in the malformation of the affected parts [4]. Treatment is usually carried out after birth when plastic and reconstructive surgery is considered to treat the resulting deformities. [4, 5]

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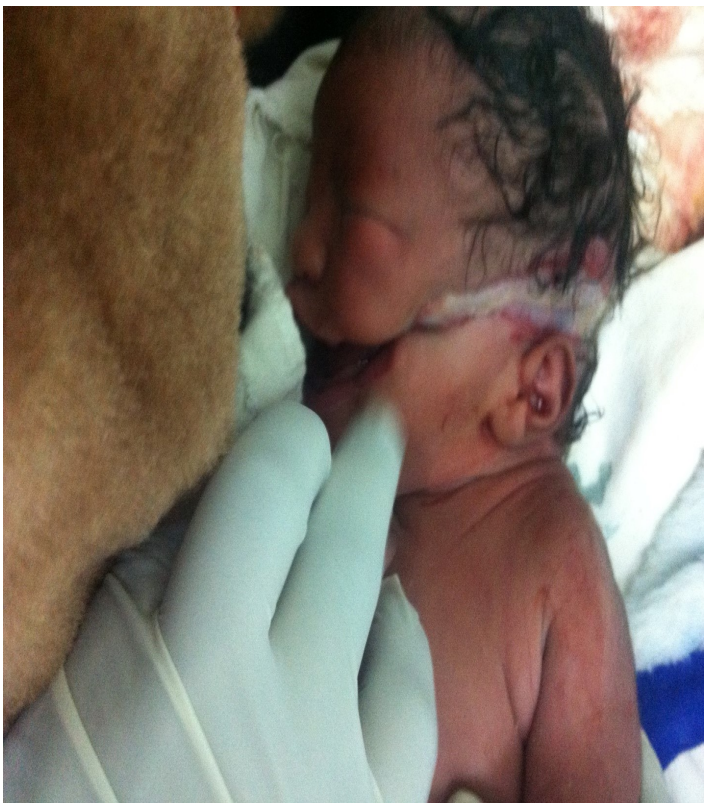
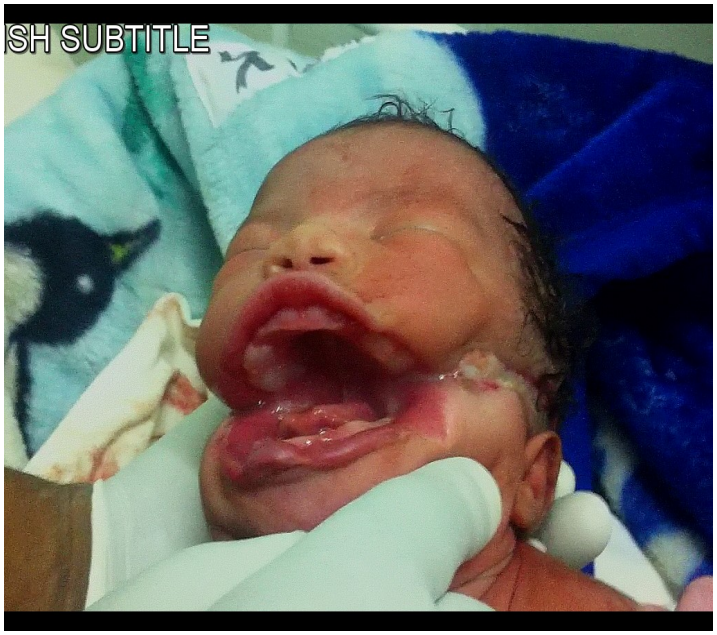
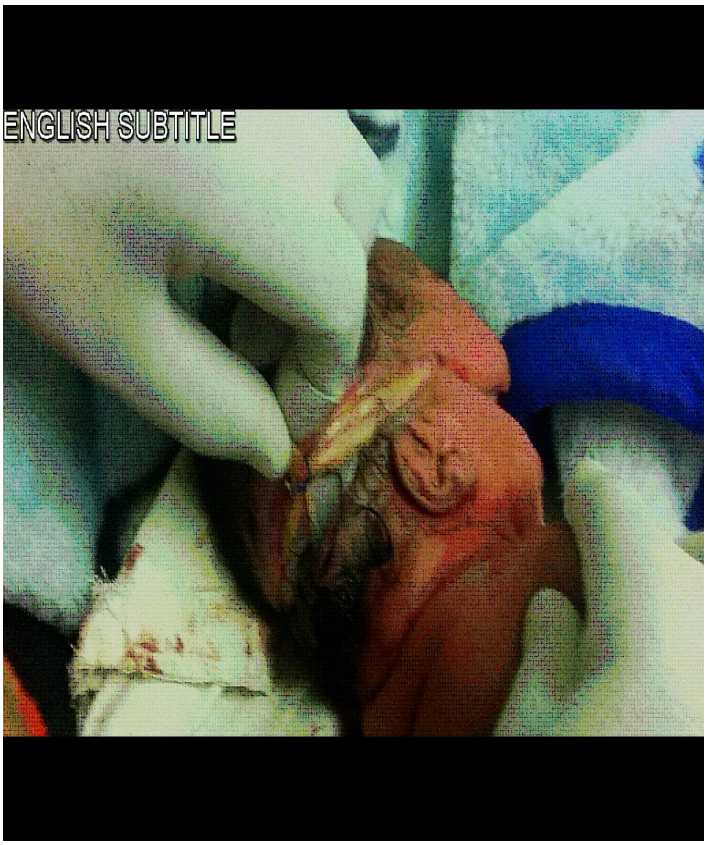
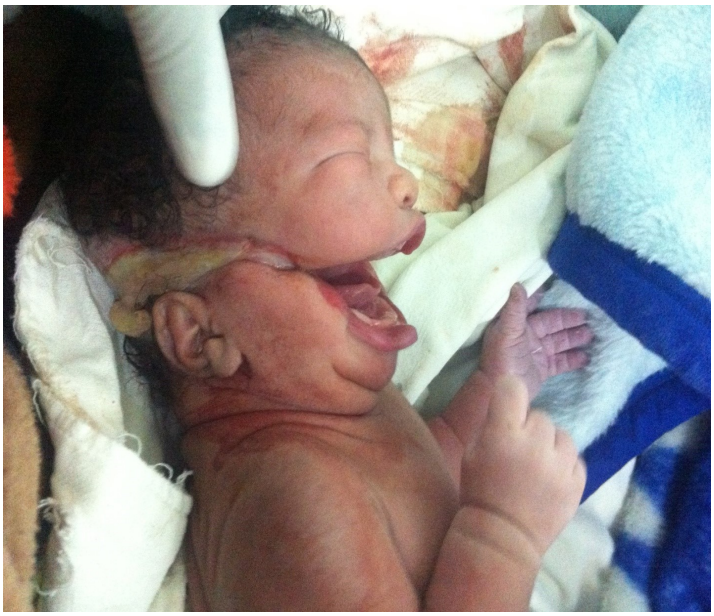
DISCUSSION

We present a case of bilateral transverse facial cleft and cranial bone defect born to a 28 year old gravida 4 para 1 mother delivered by C/S at gestational age of 41 weeks and 2 days with the indication of prolonged active first stage of labor and previous C/S scar. The pregnancy had otherwise been clinically uneventful till delivery of this female neonate weight 3120 grams with APGAR score of 8/10 in first and 9/10 in 5th minutes having stated malformations.









And the neonate cannot feed either with breast feeding or bottle due to sever bilateral transverse facial defect. On gross examination neither skeletal nor soft tissue anomalies seen except the stated cranio facial defect. X-ray of the skull shows bone defect at basal skull ,U/S examination of internal organs – normal, U/S examination of brain normal ,CT-scan of brain also showed normal finding. Neonate kept with NG-tube feeding after several attempt to have reconstructive surgery to be done for the neonate, contact made to plastic surgery and anesthesiologist was tried. After 3-4 weeks of NG feeding soft tissue defect repaired and the baby is currently in good condition and able to feed maternal breast and gaining weight with normal developmental mile stones.

Various mechanisms for the development of amniotic disruption complex have been reviewed by seeds and co-workers. Contemporary views largely favor the exogenous theory proposed by Torpin, which spontaneous rupture of the amnion leads to. ABS is a known case fetal malformations and the anomalies range from mild to sever ones that may be incompatible with post-natal life.

The exact incidence is unknown but, reported series suggest that they occur in between1 in 12,000 and 1 in 15,000 live birth and are commoner in the early pregnancy .The findings in this case are strongly consistent with ABS.

We report a case in which an initially normally formed fetus has subsequently developed an anatomic deforming attributable to this

condition. Various mechanisms for the development of amniotic disruption complex have been reviewed by seeds and co-workers[5]. Contemporary views largely favor the exogenous theory proposed by Torpin that spontaneous rupture of the amnion leads to failure of further growth of the amniotic sac. According to the extrinsic theory in the etiology of ABS, the distribution of the amnion allows the embryo or fetus to enter the chorionic cavity and contact the chorionic side of the amnion. Fetal parts may then become interrupted by the fibrous septum that traverse the chorionic space. [5]

The entanglement of fetal parts is random and the slash defects so created are non-embryologic in distribution. Fetal band and clefts could be a result of local compression or adhesion. Therefore the present case report may further support the concept of extrinsic theory in the etiology of ABS. The severity of the

band compression on the developing fetus could determine morphology of the deformity. Although severe craniofacial cleft caused by ABS are often incompatible with life, a number of these children may still survive even till school age with the accompanying facial anomaly. It has been observed that the preference of amniotic bands in consumption with craniofacial cleft is part of clinical futures of this syndrome [5, 6].

To the best of our knowledge, this is the first case to be presented in our region [ETHIOPIA].

The corrective surgical procedures for patients with ABS may range from minor to complex, and the outcome depends on the severity of the deformations [6]. The presence of a fibrous band at the base of facial cleft and scalp of this neonate suggests that ABS may be the primary cause of this defect, probably by slash effect [7]. The presented case with peculiar features of craniofacial cleft support the ideas that deformity is likely related to ABS.

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