orphanaesthesia

Anaesthesia recommendations for patients suffering from

Amniotic band syndrome

Disease name: Amniotic band syndrome

ICD 10: P02.8

Synonyms: Amniotic band constriction, ADAM complex (amniotic deformities, adhesion, mutilation), amniotic band sequence, congenital constriction bands, pseudoainhum, limb body wall complex, amniotic disruption complex, annular grooves, congenital amputation, Streeter bands, Streeter anomaly, transverse terminal defects of limb, aberrant tissue bands, amniochorionic mesoblastic fibrous strings, amniotic bands.

Disease summary: Amniotic band syndrome (ABS) consists of a wide spectrum of congenital malformations depending on the affected body part(s).

There are two hypothesis on the formation of amniotic bands and ABS. “Extrinsic model” theory explains the rupture of the amnion without the rupture of the chorion which leads to transient oligohydramnios due to loss of amniotic fluid through the initially permeable chorion. The fetus passes to the extra embryonic coelom through the defect and comes in contact with ‘sticky’ mesoderm on the chorionic surface of the amnion resulting in entanglement of fetal parts and skin abrasions. Entanglement of fetal parts by amniotic bands causes constriction rings and amputations, whereas skin abrasions can lead to disruption defects, such as cephaloceles and swallowing of the bands will cause asymmetric clefts on the face. The “intrinsic model” by Streeter suggests that the anomalies and the fibrous bands have a common origin, caused by a perturbation of developing germinal disc of the early embryo. Most cases of ABS are not of genetic origin, and occur sporadically with no recurrence in siblings or children of affected adults. Maternal trauma, teratogenic insult, oophorectomy during pregnancy, intrauterine contraceptive device, amniocentesis and familial incidence of connective tissue disorders (Ehler-Danlos syndrome) are some of the implicated etiopathological factors. [1]

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

It affects both sexes equally with an incidence of 1 in 1,200 to 15,000 live births [2] and 1 in 70 stillbirths. [3]

Due to possibility of different combinations of anomalies, there are no two identical cases of ABS. Children with ABS have very polymorphic clinical findings:

Craniofacial defects: vertical and oblique facial cleft, cleft lip and palate, orbital defects (anophthalmos, microphthalmos, enophthalmos), corneal abnormalities, microtia, central nervous system malformations (anencephaly, encephalocele, asymmetric meningocele) and calvaria defect.[4]

Truncal defects: chest wall defect with heart extrophy, lung hypoplasia, scoliosis, abdominal wall defect, abdominal organs extrophy, umbilical cord strangulation with often lethal outcome. [5]

Limb defects: constriction rings, lymphedema of the digits, shortening of the limbs or intrauterine limb amputation, amputation of the digits (most often 2nd, 3rd and 4th fingers) and toes, syndactyly, hypoplasia of the digits, club foot, pseudarthrosis, hip dislocation, peripheral nerve palsy.

Other anomalies: gastroschisis, small intestinal atresia, renal agenesis, Patau syndrome, Septo-optic dysplasia.

In 1961, Patterson described a classification [6] that is still relevant today:

a) Simple ring constrictions;

b) Ring constrictions accompanied by deformity of the distal part with or without lymphedema;

c) Ring constrictions accompanied by fusion of distal parts ranging from mild to severe acrosyndactyly and

d) Intrauterine amputations.

ABS is often difficult to diagnose before birth. Prenatal ultrasound can help in visualization of amniotic bands attached to a fetus with restriction of motion, constriction rings on extremities and irregular amputations of fingers and/or toes with terminal syndactyly. Recently 3D and 4D ultrasound techniques contribute to more sensitive prenatal diagnostics of ABS. Fetal MRI can be helpful in complicated cases. Doppler study of the constricted limb could be of useful in diagnosis of in-utero amputation as well as to take decisions regarding in-utero treatment. Physical examination is the main way of postnatal diagnosis of ABS, with a search to establish potential malformations of different organs and body parts. Ultrasound, echocardiography, and x-ray films may help to diagnose or rule out other associated anomalies.

Management strategy of ABS depends upon the extent of the associated anomalies. Treatment is mostly surgical with an individual approach to every single case. Most references recommend the use of Z-plasty or W-plasty after the excision of the constriction band, in one- or two-stage approach. Termination of pregnancy is usually proposed at the time of the diagnosis of severe craniofacial and visceral abnormalities, whereas minor limb defects can be repaired with postnatal surgery. Lately, there have been some attempts of prenatal ABS treatment – fetoscopic laser cutting of amniotic bands, before their compression on the fetus makes malformations.[7] Patterson in his study of 52 patients of congenital constriction rings

had reported only two cases of below knee amputations in addition to other musculoskeletal defects.[6] Zych, et al. in 1983 reported a case of involvement of congenital bands, pseudarthrosis and impending gangrene of leg, which was salvaged with multiple Z-plasty.[8] Greene et al. had advised a one-stage release for circumferential congenital constriction bands which was performed in all four extremities.[9] In 2006, Samra et al. reported a case of severe constricting amniotic band with a threatened lower extremity in a neonate, which was salvaged with multiple Z-plasties over a 6-year functional follow up.[10] Recently, Choulakian et al. has described a two-staged approach of direct closure after excision of the constriction band.[11] So, the outcome of the disease depends on the gravity of the malformation associated with it.

Typical surgery

Surgery is usually performed for cosmetic reasons and a staged correction may ensure the adequacy of vascularity to the residual limb or digit. Mainly release of contraction bands of the affected limbs. In-utero surgery, syndactyly, congenital amputation, 3D printing of prosthetics, cleft lip and palate repair, strabismus, clubfoot.

If the constriction is present around the digits or extremities, urgent surgical treatment may be necessary because of vascular compromise.

Type of anaesthesia

No specific recommendation for either general or regional anaesthesia. General anaesthesia along with regional anaesthesia or local anaesthesia would be a better choice in view of paediatric patients and post-operative pain management.

Necessary additional diagnostic procedures (preoperative)

A detailed history, clinical evaluation and appropriate laboratory investigations help to detect any systemic anomaly. Anaesthetic records of previous surgeries are helpful to get an idea of airway management.

ABS patients with severe scoliosis may be associated with cardiac or pulmonary diseases because of restricted thoracic size.[12] Preoperative assessment by paediatrician, cardiologist or pulmonologist should be considered in ABS patients to rule out any congenital anomalies or diseases, to assess readiness for surgery and to optimise the patient before surgery.

Particular preparation for airway management

Anaesthetists should always be prepared for difficult airway in cases with craniofacial involvement. In addition to deformities in ABS, paediatric airway is challenging because of unique anatomy (smaller size, vocal cord between C1-C4 with an anterior angulation, a large and floppy epiglottis, a large occiput) and physiology (frequent upper airway obstruction under GA, higher metabolism, and faster desaturation during period of apnoea).

Difficult airway cart with different sizes of endotracheal tubes, laryngeal mask airway, video laryngoscope, paediatric fiberscope etc. should be kept ready.

Limb reduction defects may make vascular access difficult.

Many times ABS patients are associated with cleft lip or cleft palate.[13] In patients with large cleft palate, a piece of gauge is placed to fill the gap to improve visualisation of glottis during direct laryngoscopy.

Particular preparation for transfusion or administration of blood products

The need for perioperative transfusion depends on the condition of the patient at the time of presentation. Borkar et al. reported a case of ABS in an adult female with severe anaemia (Haemoglobin 5.1 gm%) and thrombocytosis(platelet count - 785000/mm^3). Two units of whole blood was transfused to the patient.[14]

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Positioning or mobilisation of ABS patients with abnormal contracture of extremity can be difficult. So utmost care must be taken to support the limbs and pressure points should be padded with cotton rolls or gel pads appropriately prior to surgery.

Probable interaction between anaesthetic agents and patient's long term medication

Not reported.

Anaesthesiologic procedure

As cooperation is often limited in paediatric patients, sedative pre-medication and the presence of the parents during induction may be helpful.

Each patient with ABS needs an individual anaesthetic plan. The decision of Gaseous induction or intravenous induction should be based on an individual basis.

Muñoz et al reported a case of amniotic bands encompassing the umbilical cord and left lower limb which was freed by laser through fetoscopy at 21 weeks of gestation, under intramuscular anaesthesia in the fetus and epidural anaesthesia with sedation in the mother.[15] Atropine (10 µg / kg), fentanyl (15 µg / kg) and vecuronium (0.1 mg / kg) were administered intramuscularly in the fetus.

Particular or additional monitoring

To prevent hypothermia, several measure like adequate covering of extremities with warm blankets, elevation of room temperature and warm intravenous fluids can be taken. Extubation should be done when there is regular spontaneous breathing, vigorous movements of all limbs, good oxygen saturation and absence of significant hypothermia.

Possible complications

The management of this disease must be multidisciplinary and the outcome depends on the gravity of malformations.

Postoperative care

Routine post-operative care and pain management protocols should be followed.

Information about emergency-like situations / Differential diagnostics

Not reported.

Ambulatory anaesthesia

This option depends on surgical procedure, patients condition, distance between patient's home and hospital.

Obstetrical anaesthesia

None.

Literature and internet links

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Please note that that guideline has not been peer-reviewed by an anaesthesiologist, but by two disease experts instead.