Aktiv Druck & Verlag GmbH | ISSN 0170 - 5334 I 02330

www.ai-online.info

60. Jahrgang | Juni 2019

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

Offizielles Organ: Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V. (DGAI)
Berufsverband Deutscher Anästhesisten e.V. (BDA)
Deutsche Akademie für Anästhesiologische Fortbildung e.V. (DAAF)
Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

Amniotic band syndrome

Arrhythmogenic right ventricular dysplasia

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 9 | 2019

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –
a common project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

A survey of until now in A&I published guidelines can be found on:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

Projektleitung
Prof. Dr. Tino Münster, MHBA
Chefarzt
Klinik für Anästhesie und operative Intensivmedizin
Krankenhaus Barmherzige Brüder Regensburg
Prüfeninger Straße 86
93049 Regensburg,
Deutschland
Tel.: 0941 369-2350
E-Mail: Tino.Muenster@barmherzige-regensburg.de

DGAI
Deutsche Gesellschaft für Anästhesiologie & Intensivmedizin
www.dgai.de

A&I
ANÄSTHESIOLOGIE & INTENSIVMEDIZIN
www.ai-online.info

orphananesthesia

Anaesthesia recommendations for

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's bands, Streeter's 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 hypotheses on the formation of amniotic bands and ABS. The “extrinsic model” theory explains the rupture of the amnion without the rupture of the chorion which leads to transient oligohydramnios due to a loss of amniotic fluid through the initially permeable chorion. The foetus passes to the extraembryonic coelom through the defect and comes in contact with ‘sticky’ mesoderm on the chorionic surface of the amnion resulting in entanglement of foetal parts and skin abrasions. Entanglement of foetal 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 (Ehlers-Danlos syndrome) are some of the implicated aetiopathological factors [1].

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

▶ **Citation:** Mistry T: Amniotic band syndrome. Anästh Intensivmed 2019;60:S421–S427.
DOI: 10.19224/ai2019.S421

Disease summary

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 calvarial defect [4].

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

Limb defects: constriction rings, lymphoedema 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 lymphoedema;

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 visualisation of amniotic bands attached to a foetus 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. Foetal MRI can be helpful in complicated cases. Doppler study of the constricted limb could be of use in the 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.

The management strategy of ABS depends on 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 a one- or two-stage approach. Termination of pregnancy is usually proposed at the time when severe craniofacial and visceral abnormalities are diagnosed, whereas minor limb defects can be repaired with postnatal surgery. Lately, there have been some attempts of

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

prenatal ABS treatment – foetoscopic laser cutting of amniotic bands, before their compression on the foetus makes malformations [7]. Patterson in his study of 52 patients with congenital constriction rings had reported only two cases of below-knee amputations in addition to other musculoskeletal defects [6]. In 1983, Zych et al. 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 pre-operative testing (beside standard care)

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]. Pre-operative assessment by a 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, the paediatric airway is challenging because of its unique anatomy (smaller size, vocal cord between C1–C4 with an anterior angulation, a

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

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 – 785,000/mm^3). Two units of whole blood were transfused to the patient [14].

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transportation and mobilisation

Positioning or mobilisation of ABS patients with abnormal contracture of extremity can be difficult. Consequently, 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.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic 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 foetoscopic laser surgery at 21 weeks of gestation, under intramuscular anaesthesia in the foetus and epidural anaesthesia with sedation in the mother

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

[15]. Atropine (10 µg/kg), fentanyl (15 µg/kg) and vecuronium (0.1 mg/kg) were administered intramuscularly in the foetus.

Particular or additional monitoring

To prevent hypothermia, several measures 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 severity of malformations.

Post-operative care

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

Disease-related acute problems and effect on anaesthesia and recovery

Not reported.

Ambulatory anaesthesia

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

Obstetrical anaesthesia

None.

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

References

1. Mistry T, Mathur R, Saini N, Rathore P. Perioperative management of amniotic band syndrome: a case report and literature review. Anaesth Pain & Intensive Care 2015;19:505–509
2. Stevenson RE, Hall JG. Human Malformations and related anomalies, 2nd ed. New York: Oxford University Press 2006;871
3. Kalousek DK, Bamforth S. Amnion rupture sequence in previable fetuses. Am J Med Genet 1988;31:63
4. Bouguila J, Ben Khoud N, Ghrissi A, Bellalah Z, Belghith A, Landolsi E, et al. Amniotic band syndrome and facial malformations. Rev Stomatol Chir Maxillofac 2007;108:526–529
5. Poeuf B, Samson P, Magalon G. Amniotic band syndrome. Chir Main 2008;S136–S147
6. Patterson TJ. Congenital ring-constrictions. Br J Plast Surg 1961;14:1–31
7. Quintero RA, Morales WJ, Phillips J, Kalter CS, Angel JL. In utero lysis of amniotic bands. Ultrasound Obstet Gynecol 1997;10:316–320
8. Zych GA, Ballard A. Constriction band causing pseudarthrosis and impending gangrene of the leg. A case report with successful treatment. J Bone Joint Surg 1983;65A:410–412
9. Greene WB. One stage release of congenital constriction bands. J Bone Joint Surg Am 1993;75:650–665
10. Samra S, Samra AH. Threatened lower extremity in a neonate from a severely constricting amniotic band. Ann Plast Surg 2006;57:569–572
11. Choulakian MY, Williams HB. Surgical correction of congenital constriction band syndrome in children: Replacing Z-plasty with direct closure. Can J Plast Surg 2008;16:221–223
12. Laub D. Congenital Anomalies of the Upper Extremity: Etiology and Management. 1st ed., New York: Springer 2014;42–43
13. Buccoliero AM, Castiglione F, Garbini F, Moncini D, Lapi E, Agostini E, et al. Amniotic Band Syndrome: a case report. Pathologica 2011;103:11–13
14. Borkar MS, Gajbhare P, Pandey VR, Patil S. A Case of Congenital Amniotic Band Syndrome Involving All the Four Limbs With Severe Anemia With Thrombocytosis: A Rare Occurrence in an Elderly Adult. Indian J Appl Res 2015;5:24–25
15. Muñoz C, Munar F, Manrique S, Higueras T. Liberación de brida amniótica: implicaciones anestésicas. Amniotic bands division: anesthetic implications. Med Clin (Barc) 2008;131:796–797.

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

Date last modified: July 2017

This recommendation was prepared by:

Author

Tuhin Mistry, MD, Anaesthesiologist, Room No. A-92, RD Hostel, SMS Medical College and Hospital, Jaipur, Rajasthan, India
dr.tuhin2014@gmail.com

Disclosure(s) The author has no financial or other competing interest to disclose. This recommendation was unfunded.

This recommendation was reviewed by:

Reviewer 1

Anupam Das, Department of Dermatology, Building - "Prerana" 19, Phoolbagan, Kolkata, West Bengal, India
anupamdasdr@gmail.com

Reviewer 2

Sinem Ciloglu, Department of Plastic and Reconstructive Surgery, Haydarpasa Training and Research Hospital, Haydarpasa Egitim ve Arastirma Hastanesi, Plastik ve Rekonstruktif Cerrahi Klinigi, Istanbul, Turkey
eroglusinem@yahoo.com

Disclosure(s) The reviewers have no financial or other competing interest to disclose.

Please note that this recommendation was not peer-reviewed by an anaesthesiologist but by two other disease experts instead.

© Anästh Intensivmed 2019;60:S421–S427 Aktiv Druck & Verlag GmbH

Herausgeber

DGAI
Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V.
Präsident: Prof. Dr. R. Rossaint, Aachen

BDA
Berufsverband Deutscher Anästhesisten e.V.
Präsident: Prof. Dr. G. Geldner, Ludwigsburg

DAAF
Deutsche Akademie für Anästhesiologische Fortbildung e.V.
Präsident: Prof. Dr. F. Wappler, Köln

Korrespondenzadresse: Roritzerstraße 27 | 90419 Nürnberg | Deutschland
Tel.: 0911 9337812 | Fax: 0911 3938195
E-Mail: anaesth.intensivmed@dgai-ev.de

Verlag & Druckerei

Aktiv Druck & Verlag GmbH
An der Lohwiese 36 |
97500 Ebelsbach | Deutschland
www.aktiv-druck.de

Geschäftsführung
Wolfgang Schröder | Jan Schröder | Nadja Schwarz
Tel.: 09522 943560 | Fax: 09522 943567
E-Mail: info@aktiv-druck.de

Anzeigen | Vertrieb
Pia Engelhardt
Tel.: 09522 943570 | Fax: 09522 943577
E-Mail: anzeigen@aktiv-druck.de

Verlagsrepräsentanz
Jürgen Distler
Roritzerstraße 27, 90419 Nürnberg
Tel.: 0171 9432534 | Fax: 0911 3938195
E-Mail: jdistler@bda-ev.de

Herstellung | Gestaltung
Manfred Wuttke | Stefanie Triebert
Tel.: 09522 943571 | Fax: 09522 943577
E-Mail: ai@aktiv-druck.de

Titelbild
Dipl.-Designerin Monique Minde, Nürnberg

Erscheinungsweise 2019
Der 60. Jahrgang erscheint jeweils zum Monatsanfang, Heft 7/8 als Doppelausgabe.

Bezugspreise (inkl. Versandkosten):

• **Einzelhefte**	30,- €
• **Jahresabonnement:**	
Europa (ohne Schweiz) (inkl. 7 % MwSt.)	258,- €
Schweiz	266,- €
Rest der Welt	241,- €
Mitarbeiter aus Pflege, Labor, Studenten und Auszubildende (bei Vorlage eines entsprechenden Nachweises)	
Europa (ohne Schweiz) (inkl. 7 % MwSt.)	94,- €
Schweiz	90,- €
Rest der Welt	94,- €

Für Mitglieder der DGAI und/oder des BDA ist der Bezug der Zeitschrift im Mitgliedsbeitrag enthalten.

Allgemeine Geschäfts- und Lieferbedingungen
Die allgemeinen Geschäfts- und Lieferbedingungen entnehmen Sie bitte dem Impressum auf www.ai-online.info

Indexed in **Current Contents®/Clinical Medicine, EMBASE/Excerpta Medica; Medical Documentation Service; Research Alert; Sci Search; SUBIS Current Awareness in Biomedicine; VINITI: Russian Academy of Science.**

Die Beiträge aus der A&I finden Sie online unter: **www.ai-online.info**

CONTACT US

Please do not hesitate to contact us. We will be glad to answer and provide further information to you at any time.

.....
Name

.....
First Name

.....
Department / Hospital

.....
Place

.....
Telephone

.....
E-Mail

.....
Date / Signature

Please contact me for further information ○

I would like to participate in the project ○

ADDRESS

German Society of Anaesthesiology and
Intensive Care Medicine
Ursula Homberg
Roritzerstrasse 27 | 90419 Nuremberg | Germany
Tel.: +49-911-9337828 | Fax: +49-911-3938195
Email: uhomberg@orphananesthesia.eu