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

www.ai-online.info

67. Jahrgang | Juli / August 2026

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

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

Beals Syndrome

orphan anesthesia

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

SUPPLEMENT NR. 11 | 2026

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 Patientinnen und Patienten mit seltenen Erkrankungen. Damit will OrphanAnesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patientinnen und 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 Anästhesistinnen und Anästhesisten damit keine Erfahrungen gesammelt haben, sodass 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 eine Anästhesistin bzw. ein Anästhesist sowie eine weitere Krankheitsexpertin bzw. ein weiterer Krankheitsexperte (z. B. Pädiaterin bzw. Pädiater oder Neurologin bzw. Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, sodass 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 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.

Find a survey of the recommendations published until now on:

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

Projektleitung

Dr. med. Christine Gaik, DESAIC, FESAIC
Oberärztin
Klinik für Anästhesie und Intensivtherapie
Universitätsklinikum Gießen und Marburg GmbH,
Standort Marburg
Baldingerstraße
35033 Marburg, Deutschland
E-Mail:
gaikc@med.uni-marburg.de

Priv.-Doz. Dr. med. Philipp Gude, MHBA
Geschäftsführender Oberarzt
Klinik für Anästhesiologie und Intensivmedizin
Universitätsklinikum
St. Elisabeth-Hospital
Bleichstraße 15
44787 Bochum, Deutschland
E-Mail:
philipp.gude@ruhr-uni-bochum.de

DGAI
Deutsche Gesellschaft für Anästhesiologie & Intensivmedizin

www.dgai.de

A&I
ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

www.ai-online.info

orphananesthesia

Anesthesia recommendations for

Beals Syndrome

Disease name: Beals Syndrome

ICD 10: Q68.8

Synonyms: Congenital contractual arachnodactyly (CCA), Beals syndrome, Beals-Hecht syndrome, Arthrogryposis distal type 9 (DA9)

Disease summary: Beals syndrome was first described by Beals and Hecht in 1971 [1].

Beals syndrome is an extremely rare connective tissue disorder, characterized by multiple flexion contractures, arachnodactyly, severe kyphoscoliosis, abnormal pinnae and muscular hypoplasia [2,3]. The clinical features are similar to Marfan's syndrome. It differs from Marfan's syndrome in that the incidence of cardiac abnormalities like aortic root dilatation are much lower in Beals syndrome and the presence of multiple flexion contractures is characteristic of Beals syndrome. However, patients with Beals syndrome may present with mitral valve prolapse and other congenital heart disease.

Beals syndrome is an autosomal dominant condition associated with mutation in FBN2 gene on chromosome region 5q23. The incidence of Beals syndrome is unknown, and prevalence is difficult to estimate due to the overlap in phenotype with Marfan's syndrome [4]. Males and females are equally affected. Individuals with Beals syndrome are expected to be cognitively normal. Delay in the motor development is common due to contractures.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

▶ **Citation:** Sathyaseelan Ratnamma V, Smith J: Beals Syndrome. Anästh Intensivmed 2026;67:S140–S147. DOI: 10.19224/ai2026.S140

Emergency information

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Anticipate difficult airway and plan for advanced airway techniques including videolaryngoscopy and fiberoptic scopes
B	**BLOOD PRODUCTS (COAGULATION)**	No specific requirements.
C	**CIRCULATION**	Cardiac abnormalities may be present, and echocardiography should be performed.
D	**DRUGS**	No specific drugs to avoid.
E	**EQUIPMENT**	Cardiac monitoring recommended in cases of congenital heart disease.

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

Typical surgery

Scoliosis correction surgery, contracture release, reduction of long bone fractures.

Type of anesthesia

There is no definite recommendation for general anesthesia or regional anesthesia.

Beals syndrome is associated with difficult intravenous access, difficult airway and difficult positioning due to multiple contractures. There are reported cases of Beals syndrome children with difficult laryngoscopy and intubation, due to dysmorphic features [5-8].

The presence of scoliosis and / or kyphosis can present a significant technical challenge. Regional anesthesia may be challenging due to contractures and difficulties in positioning.

Necessary additional pre-operative testing (beside standard care)

Respiratory function should be assessed preoperatively as persons with Beals syndrome can have restrictive lung disease.

Children with Beals syndrome can present with various heart defects such as septal defects (ASD, VSD), interrupted aortic arch and mitral valve prolapse. A preoperative echocardiogram should be done to rule out the presence of cardiac defects and its effects.

Although ocular involvement is yet unclear, a thorough ophthalmologic evaluation is recommended.

Particular preparation for airway management

There are reported cases of difficult airway in children with Beals syndrome [5-8]. Difficult laryngoscopy and intubation are reported due to restricted mouth opening, micrognathia and high arched palate. A thorough preoperative evaluation of the airway and an adequate management plan should be in place before anesthetizing these children. Preparations for difficult airway management are advisable ranging from simple (oropharyngeal/ nasopharyngeal airways) to the advanced (video laryngoscope / fiberoptic bronchoscope).

Particular preparation for transfusion or administration of blood products

There is no evidence for specific transfusion practices in children with Beals syndrome. As in any scoliosis correction surgery, excessive bleeding and need for blood transfusion should be anticipated in children undergoing scoliosis correction surgery and general measures such as tranexamic acid and cell salvage should be considered.

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

Particular preparation for anticoagulation

No information on specific disease related pathophysiology.

Particular precautions for positioning, transportation and mobilization

Children with Beals syndrome can have multiple flexion contractures that can make positioning difficult. Special care should be taken while positioning and adequate padding of all bony protuber ances should be ensured.

Interactions of chronic disease and anesthesia medications

Not reported.

Anesthetic procedure

Both inhalational and TIVA techniques may be used. There is no evidence favoring any particular induction or maintenance agent. There are reported cases of difficult intubation in children with Beals syndrome. So a thorough preoperative evaluation of airway and an ad equate management plan should be in place before anesthetizing these children.

Regional anesthesia can be challenging due to multiple joint contractures, scoliosis and spine deformities. Additionally, scarring of the epidural space due to previous back surgery can cause unpredictable local anesthetic spreading in the epidural space, resulting in patchy, unilateral, or different-than-expected levels of the block and even reduced analgesic efficacy [9].

Particular or additional monitoring

Intraoperative monitoring needs to be tailored to the procedure and presence of comorbidities. Consider cardiac monitoring in children with cardiac comorbidities.

Possible complications

Airway issues secondary to difficulty with intubation.

Respiratory complications due to concurrent restrictive lung disease and poor airway control.

Arrythmias and hemodynamic instability can occur due to underlying cardiac abnormalities.

Injury, pressure sores and nerve damage can occur when positioning patients with muscle contractures.

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

Post-operative care

Depending on presence of comorbidities (congenital heart disease) and type of surgery performed (scoliosis correction); patients may require higher level support and monitoring in HDU/IMC or ICU postoperatively.

Disease-related acute problems and effect on anesthesia and recovery

Airway problems as mentioned above.

Ambulatory anesthesia

Minor procedures especially in patients without comorbidities can be carried out as day case procedures.

Obstetric anesthesia

Ideally maternity patients with Beals Syndrome should be referred to a tertiary unit. Neuraxial techniques are possible but may be challenging due to severe scoliosis and scarring from previous surgery [9].

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

References

1. Scola RH, Werneck LC, Iwamoto FM, et al. Congenital contractural arachnodactyly with neurogenic muscular atrophy. Arq Neuropsiquiatr 2001;59(2-A):259-262.
2. Tunçbilek E, Alanay Y. Congenital contractural arachnodactyly (Beals Syndrome). Orphanet Journal of Rare Diseases 2006;1:20. DOI: https://doi.org/10.1186/1750-1172-1-20.
3. Kyaw P, Arachchi A, Vasudevan A. A rare presentation of Beals syndrome; a newly recog nized connective tissue disorder. Grand Rounds Vol 11:115-118; Specialities: Rheumatology Article Type: Case Report. DOI: 10.1102/1470-5206.2011.0027 2011 e-MED.
4. Adam MP, Ardinger HH, Pagon RA, et al. Congenital Contractural Arachnodactyly. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2017.
5. Nagata O, Tateoka A, Shiro R, et al. Case report Anaesthetic management of two paediatric patients with Hecht–Beals syndrome. Paediatr Anaesth 1999 9: 444-447.
6. Nasreen F, Khalid A. An infant with Beals-Hecht syndrome : An airway challenge for the anaesthesiologist. Sri Lankan Journal of Anaesthesia. 2020.
7. Michalek P, Hodgkinson P, Donaldson W. Fiberoptic intubation through an i-gel supraglottic airway in two patients with predicted difficult airway and intellectual disability. Anesth Analg 2008;106(5);1501-1504. DOI: http://dx.doi.org/10.1213/ane.0b013e31816f22f6. PMid: 18420867.
8. Vazquez-Colon and Lee.Open wide: Anesthetic management of a child with Hecht-Beals syndrome. Saudi Journal of Anaesthesia. 2021.
9. Laranjeira J et al. Labor Analgesia in a Patient with Beals Syndrome : A Case Report of Management Challenges. 2025.
10. Meena, Jagdish P, Gupta, Ajay, Mishra, Devendra et al. Beals-Hecht syndrome (congenital contractural arachnodactyly) with additional craniospinal abnormality: a case report. J Paediatr Orthopaed B: May 2015;24;3:226-229.
11. Jaman NB, Al-Sayegh A. Seizures as an Atypical Feature of Beal's Syndrome. Sultan Qaboos Univ Med J 2016 Aug;16;3: e375–e378 1.
12. Gupta B. Congenital Contractural Arachnodactyly (Beals Syndrome): First Case Report with Hypospadias. Indian Paediatr 2002;39:1159-1161.

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

Date last modified: **November 2025**

This recommendation was prepared by:

Authors
Vineetha Sathyaseelan Ratnamma, Anaesthetic Registrar, Great Ormond Street Hospital, London, UK
Vineetha.SathyaseelanRatnamma@gosh.nhs.uk

Jonathan Smith, Consultant Anaesthetist, Great Ormond Street Hospital, London, UK

Disclosure: The authors have no financial or other competing interest to disclose. This recommendation was unfunded.

Peer revision 1
Lowri Bowen, Paediatric anaesthesiologist, Cardiff and Vale UHB
Lowri.Bowen@wales.nhs.uk

Peer revision 2
Jagdeep S. Walia, Head, Division of Genetics (Department of Pediatrics), Kingston, ON, Canada
jagdeep.walia@kingstonhsc.ca

Disclosure: The reviewers have no financial or other competing interest to disclose.

Update and revision (November 2025)

This recommendation was prepared by:

Authors
Vineetha Sathyaseelan Ratnamma, Anaesthetic Registrar, Great Ormond Street Hospital, London, UK
Vineetha.SathyaseelanRatnamma@gosh.nhs.uk

Jonathan Smith, Consultant Anaesthetist, Great Ormond Street Hospital, London, UK

David Cunningham, Anaesthetic Fellow, Great Ormond Street Hospital, London, UK
David.Cunningham@gosh.nhs.uk

Disclosure: The authors have no financial or other competing interest to disclose. This recommendation was unfunded.

This recommendation was reviewed by:

Review
Tino Münster, Specialist in anaesthesia, Barmherzige Brüder Hospital Regensburg, GERMANY
Tino.Muenster@barmherzige-regensburg.de

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

orphananesthesia
a project of the German Society of Anaesthesiology and Intensive Care Medicine
www.orphananesthesia.eu

Editorial Review

Christine Gaik, Specialist in anaesthesia and intensive care medicine, University Hospital Marburg, Germany
gaikc@med.uni-marburg.de

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

© Anästh Intensivmed 2026;67:S140–S147 Aktiv Druck & Verlag GmbH

Herausgeber

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

BDA
Berufsverband Deutscher Anästhesistinnen und Anästhesisten e. V.
Präsidentin: Prof. Dr. G. Beck, Mannheim

Schriftleitung
Präsident/in der Herausgeberverbände

Gesamtschriftleiter/Editor-in-Chief:
Prof. Dr. Dr. Kai Zacharowski, ML FRCA FESAIC, Frankfurt

Stellvertretender Gesamtschriftleiter/Deputy Editor:
Prof. Dr. T. Volk, Homburg/Saar

CME-Schriftleiter/CME-Editor:
Prof. Dr. W. Zink, Ludwigshafen

Redaktion/Editorial Staff
Korrespondenzadresse:
Neuwieder Straße 9 | 90411 Nürnberg | Deutschland | Tel.: 0911 9337812
E-Mail: redaktion@ai-online.info

Verlag & Druckerei

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

Geschäftsführung
Wolfgang Schröder | Jan Schröder |
Tel.: 09522 9435-0 | Fax: 09522 9435-67
E-Mail: info@aktiv-druck.de

Anzeigen | Vertrieb
Pia Müller | Tel.: 09522 9435-70
E-Mail: anzeigen@aktiv-druck.de

Verlagsrepräsentanz
Jürgen Distler
Neuwieder Straße 9 | 90411 Nürnberg
Tel.: 0171 9432534
E-Mail: jdistler@bda-ev.de

Herstellung | Gestaltung
Robert Kux | Tel.: 09522 9435-71
E-Mail: ai@aktiv-druck.de

Titelbild
Gestaltung: Roland Mehl
3D Arts GmbH
Dunantstraße 2 | 92224 Amberg
E-Mail: rmehl@3darts.de
www.3darts.de

Erscheinungsweise 2026
Der 67. Jahrgang erscheint 6x pro Jahr alle zwei Monate, beginnend ab Februar.

Die A&I ist die Verbandszeitschrift von DGAI und BDA.

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

Wichtiger Hinweis
Für Angaben über Dosierungsanweisungen und Applikationsformen kann vom Verlag und den Herausgebern keine Gewähr übernommen werden. Derartige Angaben müssen vom jeweiligen Anwender im Einzelfall anhand anderer Literaturstellen auf ihre Richtigkeit überprüft werden. Gleiches gilt für berufs- und verbandspolitische Stellungnahmen und Empfehlungen.

Der Verlag veröffentlicht die Beiträge in der von den Autorinnen und Autoren gewählten Genderform.

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
Neuwieder Straße 9 | 90411 Nuremberg | Germany
Tel.: +49-911-933780
Email: info@orphananesthesia.eu