orphanaesthesia

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

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.

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.

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.

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].

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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

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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

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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

Editorial Review

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

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