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Review Article

Difficult airway management in children and young adults with arthrogryposis

Glenn Isaacson [a,c,*], Elizabeth T. Drum [b]

[a] *Department of Otolaryngology – Head & Neck Surgery, Lewis Katz School of Medicine at Temple University, Philadelphia, PA, USA*
[b] *Department of Anesthesiology and Critical Care, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA*
[c] *Department of Pediatrics, Lewis Katz School of Medicine at Temple University, Philadelphia, PA, USA*

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Abstract *Objective:* To review current evidence and experience with anesthesia and airway management issues in children and young adults with arthrogryposis.
Data sources: Review of existing world literature and description of personal experience at a center for children's orthopedic surgery and rehabilitation over 2 decades.
Methods: Description of common problems and their solutions in this unusual and diverse group of patients.
Results: Arthrogryposis multiplex congenital includes more than 400 conditions that lead to congenital joint contractures affecting more than one body area. Among the many causes of arthrogryposis, 50%–65% fall into two large categories – amyoplasia and distal arthrogryposis. There is general agreement that best function in children with arthrogryposis is achieved through early mobilization of joint contractures. Children with arthrogryposis average >5 operative procedures during childhood. Anesthesia for these procedures may be complicated by limited jaw mobility and mouth opening, restricted lung development, positioning difficulties, difficult venous access and concerns about increased risk for malignant hyperthermia. 75% of arthrogryposis patients do not have a difficult airway. For those with a history of airway problems or those meeting criteria for a difficult airway, careful advanced planning helps to assure safe and successful surgery. We describe several specialized techniques for endotracheal intubation of children with arthrogryposis.

* Corresponding author. Department of Otolaryngology – Head & Neck Surgery, Lewis Katz School of Medicine at Temple University, 1077 Rydal Road, Suite 201, Rydal, PA 19046, USA.
E-mail address: glenn.isaacson@temple.edu (G. Isaacson).
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Conclusions: Children and young adults with arthrogryposis are a diverse group. Many pose unique challenges for airway and surgical management. Review of individual anesthesia records and careful advanced planning by a coordinated, experienced airway team can lead to best outcomes from arthrogryposis surgery.

Introduction

What is arthrogryposis?

Arthrogryposis multiplex congenital includes more than 400 conditions that lead to congenital joint contractures affecting more than one body area.[1] These conditions share the phenomena of fetal akinesia—the inability to move articulations in utero — a requirement for normal joint formation and function. The lack of mobility is associated with the development of connective tissue around the joints, which leads to fibrosis and contractures of the affected joints. Neurological diseases, muscular and connective tissue abnormalities, limited intrauterine space, in adequate placental supply, and maternal disease and infections may contribute to fetal akinesia.[2]

One hundred five different genetic defects have been identified that lead to the various arthrogryposis phenotypes.[3] Among the many causes of arthrogryposis, 50%—65% fall into two large categories — amyoplasia and distal arthrogryposis. Amyoplasia has a frequency of about 1/10,000 live births and has no genetic predisposition. It presents in the newborn period with symmetric contractures involving all 4 limbs. The jaw and trunk are relatively spared. Normal limb muscle tissue is replaced by fatty, fibrous tissue.[4]

Distal arthrogryposis is not a single entity, but a group of syndromes with joint contracture involving the hands and feet. The distal arthrogryposis syndromes affect 1/2500 live births.[5] They are subdivided into 11 types based on involvement, etiology and associated anomalies of other body structures. Freeman—Sheldon syndrome, Sheldon-Hall syndrome and Gordon syndrome are included among these 11 types. Several distal arthrogryposis syndromes have been associated with mutations in sarcomeric muscle proteins. Some include defects in embryonic myosin heavy chain protein which is expressed only during fetal life, from 6 to 24 weeks of gestational age.[6]

Management of the patient with arthrogryposis

Airway and anesthesia issues

There is general agreement that best function in children with arthrogryposis is achieved through early mobilization of joint contractures. Early therapy is so important, some have discussed early obstetrical delivery of fetuses diagnosed with arthrogryposis in utero.[7] Joint mobilization through physical therapy, casting and surgical release of contractures form the basis of infant management.[8] Children with arthrogryposis average >5 operative procedure during childhood — some many more — given the complex nature of the disorder and involvement of multiple articulations.

Anesthesia for these procedures may be complicated by limited jaw mobility and mouth opening, restricted lung development, positioning difficulties, difficult venous access and concerns about increased risk for malignant hyperthermia.[9] Several of the syndromes associated with arthrogryposis, notably Escobar (multiple pterygium) syndrome and Freeman—Sheldon (whistling face) syndrome, feature restricted jaw opening,[10] limited mouth opening, micrognathia, high-arched palate, limited cervical movement or cervical instability making direct laryngoscopy and intubation difficult.

Patients with arthrogryposis have a higher incidence of associated neuromuscular diseases or associated myopathies.[11] This has led many to be concerned about an increased risk of malignant hyperthermia. There is also frequent concern about increased incidence of intraoperative hypermetabolism and hyperthermia, although a recent large review found no evidence of increased odds of intraoperative hyperthermia.[11] In the presence of underlying myopathies, many avoid the use of succinylcholine in order to reduce the risk of hyperkalemia, although no data exists about the safety of its use in patients with arthrogryposis.[12] Non-depolarizing muscle relaxants and potent inhalational agents have an excellent safety record in anesthetic management for patients with arthrogryposis.[2]

Between 20% and 67%, of affected children have scoliosis. Repeated anesthetics are often necessary for expansion of exoskeletal spinal implants used to correct spinal alignment. Vertebral curves over 50° in individuals nearing or past skeletal maturity may require extensive, protracted spine fusion surgery.[13] These are associated with substantial blood loss and can be complicated by post-operative respiratory problems from compromised lung function, underlying myopathies and surgical stress. This should be anticipated during pre-operative planning and may affect both the timing of post-operative extubation and the need for postoperative intensive care unit facilities.

Approach to airway management

Regional anesthesia

When planning operative interventions, the first question to ask is whether general anesthesia is necessary. While this is the default choice in healthy infants and children, local and regional techniques have important advantages in children with arthrogryposis. For upper extremity surgery, axillary,

supraclavicular, and infraclavicular blocks have been used for anesthesia and for post-operative pain control. The blocks are often applied under ultrasonographic guidance. For lower extremity surgery, paravertebral, lumbar epidural, iliofascial, femoral nerve, and sciatic nerve blocks provide a similar function. The use of indwelling catheters can help control postoperative surgical pain and pain during aggressive postoperative passive range of motion therapy.[14] While successful spinal anesthesia has been reported in children with limb[15] fractures and for affected adults during caesarean section and ex utero intrapartum treatment, there are concerns given the high frequency of vertebral column deformities in arthrogryposis.[16] The use of regional anesthesia does not obviate the need to have careful planning for airway management in the event of failed or incomplete block.

Planning

Careful pre-operative planning is important when general anesthesia is chosen. As individual patients may have very different airway problems, review of anesthesia records from previous surgeries is especially important. These can help to identify problems with ventilation and laryngeal exposure. If the last airway team found a solution to this patient's difficult airway problem, it is prudent to follow a similar approach. Finally, as 75% of arthrogryposis patients do not have a difficult airway, past experience can help to guide the need for a full airway team during anesthesia induction. Our difficult airway team includes a pediatric otolaryngologist for children with arthrogryposis and suspected or established challenging airways. The otolaryngologist adds skills in rigid bronchoscopy and tracheotomy, should the standard anesthesia difficult airway equipment and maneuvers fail. Careful planning for airway management, taking into consideration the resources available in the surgical suite is paramount for patients with arthrogryposis.

The Shriner's Hospital for Children — Philadelphia, where the authors have worked over the last decades, serves as a major center for arthrogryposis surgery. In a typical year over 200 children undergo upper limb (25%) or lower limb (75%) operative procedures and 5—10 have corrective spine surgery. Our experience (and that of others[12,13])suggests that most arthrogryposis patients, including those with limitation of jaw opening and neck extension, are easily supported by mask ventilation. Thus inhalation induction with subsequent intravenous access is usually safe. The next important question is whether endotracheal intubation is required for airway support. For those children undergoing procedures on the upper or lower limbs that do not require prone positioning, ventilation by mask (for brief procedures) or via laryngeal mask airway is often sufficient.[17]

Intubation

Several specialized techniques have proven effective for endotracheal intubation of children with arthrogryposis. Anesthesiologists often recommend induction with preservation of spontaneous ventilation when approaching a potentially difficult airway for the first time.[18] While pediatric otolaryngologists may prefer the superior exposure and illumination of Parsons or Lindholm laryngoscopes,in infants and children with small mouths (e.g. Freeman—Sheldon syndrome), a standard anesthesia laryngoscope with a narrow, curved blade (Phillips or Wisconsin/Wis-Hipple) is often more effective effect.

Limited jaw opening and/or neck extension may produce the appearance of an "anterior larynx".[19] In many cases, the addition of cricoid pressure will expose at least the arytenoids and allow for intubation with a styletted endotracheal tube. Occasionally, the tip of the endotracheal tube can be advanced to the laryngeal introitus, but not beyond. This may result from the angled tube and stylet catching on the anterior cricoid ring. This obstruction may be overcome if the stylet is withdrawn slightly and the endotracheal tube is rotated 90—180°. If this fails, introduction of a bougie or endotracheal tube changer first to guide the endotracheal tube may permit intubation.[20,21]

If the larynx remains out of view with a convention laryngoscope blade, video laryngoscopes (e.g. Glidescope™, Verathon, Seattle, WA) can provide necessary exposure. Unfortunately, the thick plastic videolaryngoscope blade occupies a large portion of a small oral airway making advancement of the endotracheal tube difficult despite good visualization. In this setting, using a flexible nasopharyngoscope as a video-stylet following exposure by a conventional metal laryngoscope may prove more effective.[22] Several authors have recommended support of the airway with a supraglottic airway device. An endotracheal tube is then introduced through the supraglottic airway and advanced under flexible bronchoscopic guidance.[23,24]

When to do a tracheotomy

For children with established difficult airways who need multiple surgical procedures over a short period of time, tracheotomy is desirable. The risks of tracheotomy are outweighed by those of airway injury or hypoxia during repeated high-risk intubations. In addition, children with difficult airways and limited pulmonary reserve (from scoliosis or myopathies) may benefit from post-operative positive pressure ventilation. This is facilitated by tracheotomy as sedation and/or paralysis needs are less when orotracheal intubation is avoided. Finally, as the full airway team is seldom available at night, tracheotomy lessens the risks attendant to accidental extubation in the intensive care unit.

There is a small subset of children who cannot be intubated by any of the previously mentioned techniques. In such cases, where aborting the operative procedure is not an option, controlled tracheotomy with mask or laryngeal mask airway ventilation can be done. Even in skilled hands, the risk of operative error is increased when the trachea is not supported by an endotracheal tube.

Conclusions

Children and young adults with arthrogryposis are a diverse group. Many pose unique challenges for airway and surgical management. We have reviewed our experience at the Shriner's Hospital for Children — Philadelphia and other

centers around the world in hopes of improving the perioperative anesthesia and airway management for this complex patient group. Review of individual anesthesia records and careful advanced planning by a coordinated, experienced airway team can lead to best outcomes from arthrogryposis surgery.

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Conflicts of interest

None.

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