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# Perioperative Care of the Child with Arthrogryposis

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## INTRODUCTION

Arthrogryposis multiplex congenita (AMC) is a rare condition characterized by congenital joint contractures throughout the body which results from fetal akinesia due to extrinsic factors or primary neurogenic or myopathic conditions of the fetus.

Children with arthrogryposis often require multiple orthopedic corrective procedures. There are three major recognized groups: Amyoplasia, distal arthrogryposis and syndromic arthrogryposis, all of which present various anesthetic challenges.<sup>1</sup>

## CASE DESCRIPTION

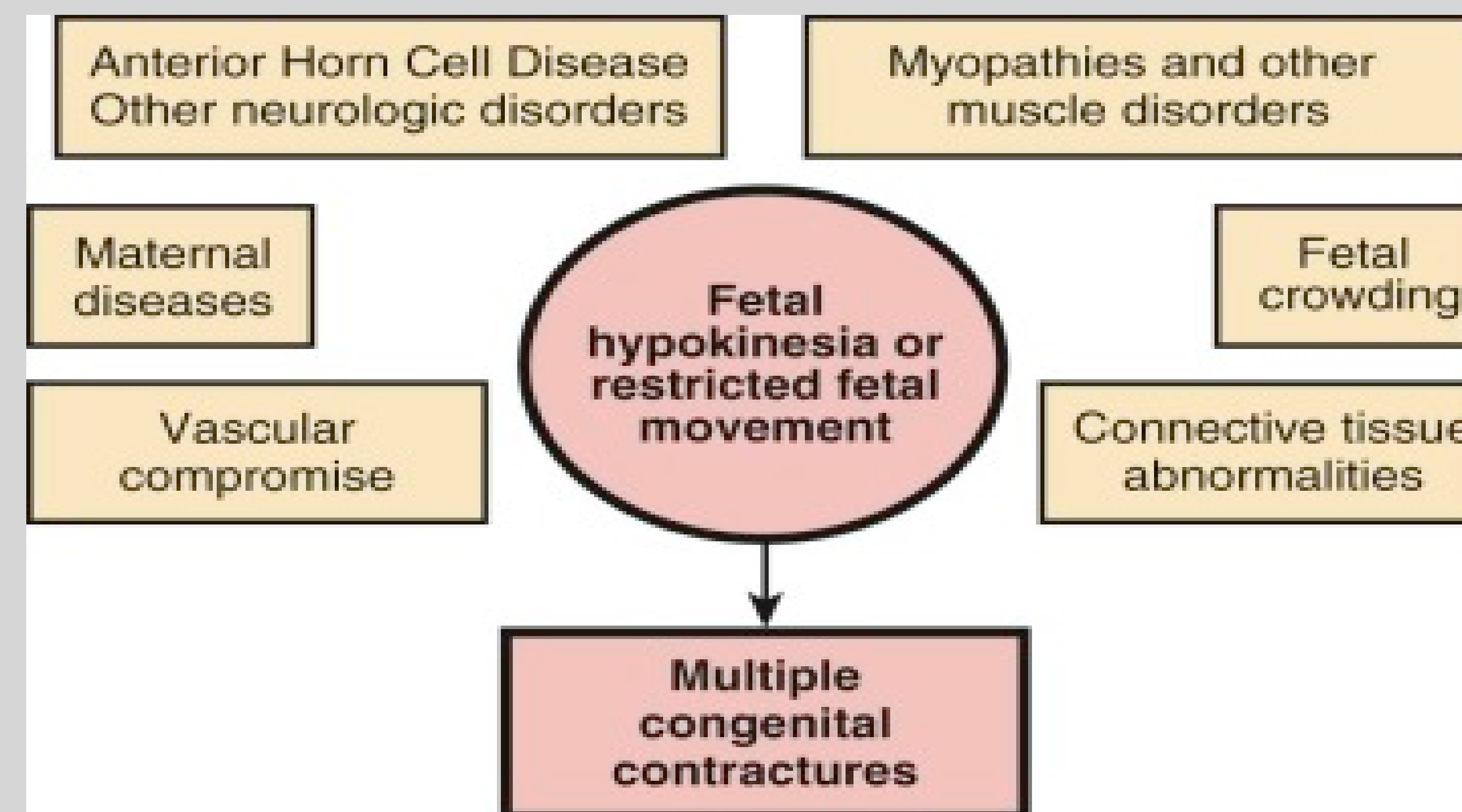
An eight year old female weighing 20kg, with distal arthrogryposis, presented for a right foot cast change and wound check. She had a history of significant right foot fixed equinus contracture; a right foot contracture release and Achilles tendon lengthening had been performed under general anesthesia a month prior to presentation.

Patient was brought into the operating room, standard monitors placed and inhalational induction performed with Oxygen/Nitrous oxide and sevoflurane. A 22G left peripheral access was obtained post-induction and the airway secured with a laryngeal mask airway size 2.5.

The anesthetic was maintained with sevoflurane and fentanyl. Intra-operatively, her vitals remained stable and nasopharyngeal temperatures were normal. She was successfully extubated in the operating room and post-operative course was uneventful.



Bilateral clubfoot deformity in a newborn with arthrogryposis. Arthrogryposis: Ferguson J. Orthopaedics and Trauma, Volume 27, Issue 3, 171-180. Copyright © 2013 by Elsevier.



Etiology of arthrogryposis. (Modified from: Hall JG. Arthrogryposis multiplex congenital: etiology, genetics, classification, diagnostic approach, and general aspects. *J Pediatr Orthop B*. 1996; 6:159–166). Arthrogryposis: Horstmann HM. Nelson Textbook of Pediatrics, Chapter 682, 3310-3314.e1. Copyright © 2016 by Elsevier.

## CASE DISCUSSION

Patients with AMC present with a wide variety of clinical features which must be taken into consideration prior to administering an anesthetic to ensure safe delivery of anesthesia. Clinical features include<sup>1,2,3</sup>:

- Difficult airway: Micrognathia, high arched palate, cervical spine instability
- Intraoperative hyperthermia: Unclear Malignant Hyperthermia risk
- Difficult IV access due to reduced subcutaneous tissue
- Difficult positioning
- Difficult regional blockade
- Myopathy: Prolonged duration of neuromuscular blockade
- Cardiorespiratory status: Sensitivity to anesthetic agents and high risk of aspiration or post-operative respiratory support needs

## CONCLUSION

*The overall prevalence of arthrogryposis multiplex congenita is estimated at 1 in 3000 live births. Anesthesiologists need to appreciate the multiple clinical presentations and anesthetic risks to help formulate an appropriate anesthetic plan for pediatric patients presenting for multiple corrective procedures.*

## REFERENCES

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