Clinical Reports

Anaesthetic management of an obstetrical patient with arthrogryposis multiplex congenita

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Arthrogryposis Multiplex Congenita (AMC) describes a rare multifactorial syndrome caused by a variety of neurogenic and myopathic disorders first diagnosed at birth and often progressing to a state of significant disability. It includes abnormalities of the cardiovascular, respiratory, nervous and genitourinary systems. The syndrome is seen primarily in the paediatric population, and orthopedic procedures are common. There remains much confusion regarding diagnosis, aetiology, genetic probabilities, natural history and management.[1-2] Occasionally an adult with this sydrome presents for an anaesthetic.

The following is a case report of a patient with the manifestations of this syndrome presenting for a primary Caesarean section.

Case report

A 21-year-old GI PO presented to her obstetrician for care during her first pregnancy. Because of her severe musculoskeletal abnormalities, including pelvic deformities, an elective Caesarean section was planned for her 38th week of gestation. Her obstetrical history was otherwise unremarkable.

The patient was well informed about her disease and had been worked up extensively elsewhere for possible multisystem involvement. She had no known associated central nervous, cardiac, urologic or primary respiratory involvement. The most significant problems were a marked kyphoscoliosis, pelvic and lower limb deformities.

Physical exam revealed a young female with a normal size thorax and greatly reduced limb length. She was confined to a wheelchair but manoeuvred it easily. She could open her mouth fully but had limited extension of the head and her neck. She had a complete set of teeth. She had almost no lateral movement of her head and neck. Her pre-pregnancy weight was 37 kg and at term she weighed 45 kg.

Significant past medical history included multiple orthopaedic operations as a child. They were done in a different country and she had no recall of any details relating to these operations; the last one had been done more than 15 years previously.

After a discussion of the possible anaesthetics for Caesarean delivery, it was decided that an epidural would be attempted.

On the morning of the operation the patient was taken to the operating room after receiving intramuscular cimetidine and oral sodium citrate. She was placed in the sitting position with ECG and BP monitors applied and her back was prepped and draped. After local skin infiltration a 16 gauge Tuohy needle was placed with difficulty at the L_{3-4} interspace and 4 ml of two per cent CO_2 lidocaine with epinephrine (5 $\mu g \cdot ml^{-1}$) was injected through the needle and a catheter was easily threaded into the epidural space to the 8 cm mark.

A total of 18 ml of local anaesthetic (two per cent carbonated lidocaine with epinephrine) was injected in 4 ml increments, over 20 minutes. A dense left-sided sensory block extending to the second thoracic dermatome developed with virtually no change in the right-sided sensation. Since the total dose of local anaesthetic was high (18 ml × 20 $mg \cdot ml^{-1}$ = 360 mg/45 kg) it was decided to proceed to a general anaesthetic. With the

Key words

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patient in the left lateral tilt position and pre-oxygenated by a tight fitting O_2 mask, a rapid-sequence induction with cricoid pressure using thiopentone (2.5 $mg \cdot kg^{-1}$) and succinylcholine (1 $mg \cdot kg^{-1}$) was started. When her peripheral twitch had disappeared, laryngoscopy was done. What was thought to be a Grade II airway[7] appeared more difficult with only the tip of the epiglottis visible (Grade III). A second attempt using a smaller endotracheal tube (6.0) and a different blade lifting the epiglottis was successful. She was maintained on N_2/O_2 (50/50) with isoflurane 0.5 per cent until delivery, then the anaesthetic was deepened to N_2O/O_2 (70/30) with 50 μg of intravenous fentanyl.

The procedure was started and a normal term baby with Apgar scores seven and nine at one and five minutes respectively was born five minutes after the incision. The rest of the operation was uneventful with no other muscle relaxant being necessary. The peripheral twitch returned 20 minutes after induction (approximately the time of skin closure).

The patient was extubated awake at the conclusion of the operation and did well postoperatively.

TABLE I Congenital anomalies associated with Arthrogryposis Multiplex Congenita

Organ system	*Abnormality*
Head and neck	Craniosynostosis
	Mandibulofacial dysostosis
	Micrognathia
	Microphthalmia
	Facial diplegia (Mobious' syndrome)
	Klippel-Feil syndrome
	Low set ears
	High arched palate
Shoulder	Sprengel's deformity
Spine	Vertebral anomalies
	Spina bifida
	Sacral agenesis
	Scoliosis – kyphosis
Cardiovascular system	Congenital heart disease
Respiratory system	Tracheoesophageal fistula
	Hypoplastic lungs
Genitourinary system	Renal anomalies
	Cryptorchidism
	Scrotal defect
	Labial defects
	Absence of the vagina/uterus
Abdomen	Inguinal hernias
Extremities	Absence of patellas
	Syndactylism
	Constriction bands
	Clinodactyly

From Thompson.[6]

Discussion

Arthrogryposis Multiplex Congenita is a symptom complex of congenital joint contractures associated with both neurologic and myopathic aetiologies. There are often accompanying developmental defects in the neurologic system as well as of the viscera.

Since first described by Otto in 1841[3] as a congenital myodystrophy, and subsequently by Stern in 1923 as AMC, it has undergone much study by the paediatric orthopaedic physicians. Its true multiplicity is only now becoming apparent with confusion regarding diagnosis, aetiology, genetic probabilities and management.

The joint deformities in AMC are primarily neurogenic (>90 per cent) in origin with myopathic causes making up the remainder.[6] Each of these groups may be divided into multiple sub-groups depending on the location of the lesion and microscopic or biochemical pathology.

Clinically, the syndrome is manifested as multiple joint contractures with onset during fetal development. Alteration takes place in the final common pathway of neuromuscular development (i.e., anterior horn cells, roots, peripheral nerves, motor end plates or muscle fibres). Intrauterine states which interfere with fetal movement may also contribute to the joint abnormalities.[8]

Visceral abnormalities (Table I) include cardiac lesions (PDA, aortic stenosis, contraction of the aorta and cyanotic heart disease), pulmonary lesions (restrictive lung disease) and central nervous system lesions (anencephaly), genitourinary problems and craniofacial deformities.

Other pathologic syndromes have been misdiagnosed as AMC at birth or later, confusing further the picture of true AMC. These have included central core disease, congenital chromosomal abnormalities and myasthenia gravis.

The anaesthetic considerations for AMC have not often been described, primarily because of the confusion in aetiology. Each differing pathophysiologic abnormality must have its own special considerations based on its unique mechanism of disease.

The physical deformities must be addressed, each deformity presenting its own problems. Airway problems, including micrognathia, cervical spine abnormalities and cervical flexion deformities, may be present. Respiratory compromise from thoracic cage abnormalities may make patients more susceptible to the respiratory depression of various intravenous and inhalation anaesthetics. Patients may be respiratory cripples from severe scoliosis prior to any corrective surgery.[10] Lumbar spine abnormalities may make correction impossible in some patients coming for other surgery (Table II).

Both neurologic and myogenic causes of AMC may cause patients to react abnormally to muscle relaxants. There

TABLE II Anaesthesia considerations in the patient with Arthrogryposis Multiplex Congenita

Anaesthetic concern	*Abnormality*
Airway	Micrognathia Cervical spine deformities High arched palate
Induction agents	Increased sensitivity secondary to decreased muscle mass
Muscle relaxants	Increased sensitivity secondary to decreased muscle mass
Succinylcholine	Increased susceptibility to MH with myogenic causes of AMC
Maintenance – inhalation agents	? susceptibility to MH with myogenic causes of AMC Increased risk of respiratory depression unless ventilation supported
Regional block	Deformities of vertebral column Abnormal spinal cords (decreased anterior horn cells) Abnormal CSF Joint contractures – Decreased accessibility to nerves

have been several reports of hyperthermia after anaesthesia in some AMC children.[4,5] Other series[9] report no cases of malignant hyperthermia, or of temperature changes during anaesthesia.

In our patient, the aetiologic mechanism of her AMC was not known. She had had multiple anaesthetics as a child without incident, but none as an adult. Making the problem a little more interesting were the anaesthetic considerations for Caesarean section. After evaluation and discussion with our patient, it was decided that a regional block would be attempted.

The possibility of abnormal spinal cord CSF dynamics, and the ease with which an epidural may be titrated to an appropriate level made the choice of epidural anaesthetic preferable to spinal anaesthesia. When a unilateral block developed it supported the possibility of abnormal development, and because of the difficulties with insertion and total dose of local anaesthetic given, it was decided against redoing the epidural.

The administration of a general anaesthetic also presented problems ascribed to AMC. A Grade II airway was anticipated so a rapid sequence induction was done after preoxygenation and cricoid pressure. The fact that it was more difficult reinforces the concern about abnormal airways and cervical spines in these patients.

Finally, her response to succinylcholine was prolonged, but not out of the range seen in some other pregnant patients.

In conclusion, we describe a patient with AMC requiring Caesarean section for delivery. A major regional block was not successful, possibly due to musculoskeletal abnormalities of the lumbar spine, and general anaesthesia was difficult due to airway abnormalities.

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Résumé

La conduite anesthésique chez une femme âgée de 21 ans atteinte d'arthrogrypose congénitale et devant subir une césarienne élective est décrite. L'anesthésie régionale fut tentée mais sans succès. L'anesthésie générale ainsi qu'une discussion des différentes considérations est présentée. La conduite anesthésique dépend surtout sur des manifestations spécifiques individuelles et les problèmes associés.