Anesthesia for Arthrogryposis Multiplex Congenita

- Report of 12 cases -

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Arthrogryposis multiplex congenita (AMC) is a clinical syndrome which includes congenital, non-progressive multiple and persistent joint contractures with generalized muscle wasting[1]. Its other associated abnormalities include skeletal deformities, congenital heart disease[2], and respiratory and genitourinary system anomalities[3,4]. Due to the severity of their joint deformities, affected patients may require multiple orthopedic procedures. They present anesthetic risk such as malignant hyperthermia[5,6], and difficulty in tracheal intubation[7]. Although various kinds of anesthetic technique[7,8], anesthetics[5,9], and muscle relaxants[9] have been recommended for these patients, each of them has both advantages and disadvantages. We performed a total of 45 anesthetic procedures in 12 patients with AMC, using mainly nitrous oxide and halothane, and assessed these patients' risk in terms of general anesthesia.

Cases

Twelve patients (8 male, 4 female), in whom AMC had been diagnosed during the period 1975 to 1986, were retrospectively studied. The patients' ages ranged from 3 months to 14 years. There was no evidence for familial incidence of AMC in these patients, except for one whose grandfather had a joint contracture suggestive of AMC. Prenatal complications of these patients included premature rupture of the membranes (1 case), coiling of the umbilical cord (2 cases), weak labor pain (3 cases) and fetal distress (2 cases). Two patients were delivered under breech presentation and two were delivered under caesarean section. Orthopedic and associated anomalities are shown in table 1. Cardiovascular anomalities were not seen among these patients. Only three patients had no AMC-associated anomalities. Preanesthetic laboratory findings revealed elevated serum creatine phosphokinase in 3 patients, elevated alkaline phosphatase in 7 patients and elevated lactate dehydrogenase in 5 patients. A severe restrictive respiratory dysfunction was noted in one patient suffering from scoliosis.

The patients underwent a total of 45 surgical procedures, mainly orthopedic procedures including release of contracture, skin graft of hands, and correction of scoliosis under general anesthesia. The patients received from 1 to 7 anesthetic procedures each. Atropine, for infants under 1 year of age, and secobarbital and atropine for other patients were intramuscularly given as premedication. In 42 surgical procedures, anesthesia was induced with halothane via face mask and maintained with nitrous oxide and halothane. Ketamine was used for a short arthrographic procedure in one pa-

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Table 1. Associated anomalities in 12 patients of arthrogryposis multiplex congenita

Anomalities	Number of patients
Talipes varus	5
Scoliosis	2
Wry neck	2
Small mandible	1
High arched palate	1
Funnel chest	1
Lordosis	1
Inguinal hernia	1
Cryptorchidism	1
Muscular atrophy of upper arm	1
Constriction band syndrome	1
Dislocation of the hip joint	1
Cerebral palsy	1

tient. Enflurane and fentanyl were used for surgical corrections of scoliosis in 2 patients, during which epinephrine was administered to decrease blood loss. Tracheal intubation was performed in 36 procedures. Suxamethonium was administered to facilitate tracheal intubation in 4 procedures, whereas deep-inhalation anesthesia without any muscle relaxant was used for tracheal intubation in the others. Tracheal intubation was difficult in 7 patients of the twelve. In a one-year-old female, intubation was extremely difficult because of her small mandible and difficulty in opening her mouth. In this case, laryngospasm, cyanosis and bradycardia developed at several trials of tracheal intubation under nitrous oxide and halothane anesthesia. Intubation was finally performed successfully after 2 hr of attemps. In a seven-year-old female, cardiac arrest occurred during anesthetic induction with nitrous oxide and halothane because her airway could not be kept patent with face mask. Rapid tracheal intubation and cardiac massage were performed successfully. She developed no post-surgical sequelae related to the cardiac arrest.

An abnormal elevation of body temperature was noticed in a one-year-old female who underwent a total or 5 anesthetic procedures. Her rectal temperature increased from 36.3°C to 38.5°C during nitrous oxide-halothane anesthesia, but she was effectively treated by surface cooling alone. Cardiac arrhythmia and muscle rigidity were not observed and blood gas data were normal during anesthesia. A 14-year-old male with severe restrictive respiratory dysfunction (vital capacity 25.6%) underwent a surgical correction of scoliosis under intravenous fentanyl and enflurane inhalation. Hypercapnea developed during anesthesia in spite of assisted ventilation, and continued several hours in the postoperative period. In a 12-year-old male, copious oropharyngeal and tracheal secretions were noted during nitrous oxide and halothane anesthesia and tracheal suctioning was frequently required. The problems which occurred in these AMC patients during anesthetic procedures are summarized in table 2. Only 3 patients (25%) had no complications during anesthesia. The postoperative course was uneventful in all 12 patients.

Table 2. Problems associated with AMC during anesthesia

Problems	Number of patients
Difficult intubation	7
Difficulties with airwary security	1
Hyperthermia	1
Respiratory acidosis	1
Copious airway secretions	1

Discussion

AMC was first described by Otto in 1841, who referred to it as congenital myodystrophy. Numerous causes of this condition have been proposed, but none has been generally accepted. Some investigators have regarded AMC as a hereditary condition[2], whereas others have not[10].

A high incidence of anomalities associated with AMC has been reported by numerous authors[2–4]. The most common anomalities are orthopedic ones such as club foot and scoliosis. The most frequent non-orthopedic anomalities occur in the genitourinary sys-

tem such as cryptorchidism[4]. The anomalities seen in our 12 patients were consistent with these reports. Two previous reports showed that about 9% of affected patients had associated congenital heart disease such as patent ductus arteriosus, aortic stenosis, coarctation of the aorta, and cyanotic heart disease[2,4]. Our patients with AMC had no cardiac anomalities.

Patients with AMC frequently require repeated surgical procedures to correct joint deformities. Spinal and epidural anesthesia have been recommended, when indicated, by some investigators[7,8]. But they may require general anesthesia, because the majority of them are under 10 years of age and surgical sites frequently include the upper extremities. Problems arising during anesthesia may include difficulties in securing the airway, in tracheal intubation due to micrognathia and short neck[9], in positioning due to multiple joint deformities, in cannulation of blood vessels due to venous maldevelopment[11] and characteristic skin shape[6], in respiratory management due to skeletal deformities such as scoliosis[9] and airway secretions, and in hemodynamic management due to congenital heart disease. In addition, a high incidence of malignant hyperthermia in these patients has been reported[11].

We performed 45 general anesthetic procedures for 12 patients with AMC. Anesthetic induction was accomplished usually without difficulty using oxygen, nitrous oxide and halothane delivered via face mask, but in one patient cardiac arrest occurred because of difficulty in securing the airway. Tracheal intubation was difficult in 7 of 12 patients. One patient who brought about extremely difficulty in tracheal intubation developed cyanosis and bradycardia.

Baines et al.[12] performed 256 anesthetic procedures for patients with AMC using halothane without an episode of malignant hyperthermia, and they concluded that the use of halothane in patients with AMC may be justified. Baines et al.[12] also used suxamethonium for 4 patients without an episode of malignant hyperthermia. We avoided intravenous administration of suxamethonium in most of our patients. Although we used suxamethonium for 4 patients to facilitate tracheal intubation, hyperthermia did not develop in any of them. However, muscular degeneration and hyperthermia associated with suxamethonium has been reported in patients with AMC[6], so we believe its use should be avoided. Elevation of body temperature occurred in one of our patients during anesthesia with nitrous oxide and halothane. We do not, however, believe malignant hyperthermia had occured, because the condition was easily treated by surface cooling and no other stigmata of malignant hyperthermia were evident.

Honda et al.[5] reported that althesin could be safely used for patients with AMC who are susceptible to malignant hyperthermia. Patients with AMC are prone to hypotension and respiratory depression due to associated myopathy and skeletal deformities. Oberoi et al.[9] recommended the use of ketamine anesthesia for a patient with AMC because of its relative lack of respiratory and cardiovascular depressive effects. On the other hand, a patient with AMC who produced copious orophangeal secretions has been reported[7]. In such a patient the use of ketamine should be avoided, because the volume of such secretions tends to be increased by ketamine. In the present study, ketamine was used in only one anesthetic procedure for an arthrography. Further study is needed to assess the safely and effectiveness of ketamine anesthesia for patients with AMC.

In summary, we performed 45 general anesthetic procedures for 12 patients with AMC (aged 3 months to 14 years) who had various orthopedic and AMC-associated anomalities. Nitrous oxide-halothane could be used safely without the increasing risk of malignant hyperthermia, but because the use of suxamethonium is still controversial, it was avoided in most of anesthetic procedures. Anesthesia should be carefully induced in patients with AMC because of potential difficulties in securing the airway and with tracheal intubation.

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