

Anesthesiology
55:597-598, 1981

Caudal Anesthesia for Early Onset Myotonic Dystrophy

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The myotonic syndromes are a group of hereditary degenerative diseases of skeletal muscle characterized by an abnormal delay in relaxation following contraction.¹ Early onset myotonic dystrophy is a rare variant in which symptoms become apparent in the neonate.²⁻³ These symptoms consist of difficulty in sucking and swallowing, facial diplegia, generalized muscle hypotonia, talipes equinovarus, and a delay of psychomotor and language development. This report describes the use of regional anesthesia for an infant who had early onset myotonic dystrophy.

REPORT OF A CASE

A two-year-old girl, weighing 8.9 kg was scheduled for bilateral repair of a talipes equinovarus deformity. Maternal family history was strong for grip myotonia. At birth, delivery was complicated with Apgar scores of three at one minute and seven at five minutes and associated meconium staining. At two days of age, hypotonia in both lower extremities, facial diplegia, and bilateral talipes equinovarus deformities were noted. Later sucking and swallowing difficulties and delay of language development occurred. The electromyogram was characterized by spontaneous high-frequency discharges which were compatible with early onset myotonic dystrophy and computerized axial tomography showed that the patient had hydrocephalus with cortical atrophy. The current physical examination revealed a small floppy child with poor head control and decreased extremity strength especially in the lower limbs. Physical examination and laboratory values were otherwise unremarkable.

Meperidine, 12 mg, promethazine, 3.1 mg, and chlorpromazine, 3.1 mg, were given orally one hour prior to surgery. Anesthesia was induced by inhalation 70 per cent nitrous oxide and 0.5 per cent halothane. Once the eyelid reflex had disappeared, the patient was turned to the later position. The skin over the sacrum was cleansed and the sacral cornua were identified. A 22-gauge 2.5-cm needle was inserted through the skin overlying the sacral hiatus. The needle was positioned at an angle of 65-70° to the horizontal, in an upward direction, with the bevel towards the feet. On piercing the sacrococcygeal ligament, there was a characteristic yield until the needle point was arrested by the anterior table of the sacrum. This was taken as a sign that the extradural space had been entered. As distinct from the technique in adults, the needle was not advanced in the sacral canal more than 0.5 cm, thus puncturing the dural sac was avoided (the distance between

the sacral hiatus and the dural sac may be very small, as little as 1 cm at this age). A small amount of air (0.2 to 0.3 ml) was carefully injected to assure that the needle point was anterior to the posterior table of the sacrum. Following a negative aspiration test for blood and cerebrospinal fluid, a test dose of 2 ml 0.5 per cent bupivacaine was injected. After waiting for three minutes when no signs of hypotension or convulsion were apparent, 6 ml 0.5 per cent bupivacaine was injected.

The child was then placed in the supine position and the halothane was discontinued. The onset of caudal anesthesia was evident within five minutes of injection by an increase in temperature in both lower extremities, loss of muscle tone, and relaxation of anal sphincter. Withdrawal of the lower limbs upon application of an electrical stimulus from a nerve stimulator in the unblocked area indicated that the block extended to the umbilicus. The operating table was kept horizontal throughout the procedure which began 15 minutes after placement of the caudal anesthetic.

The intraoperative course was uneventful and total anesthesia time was 150 minutes. The degree of muscle relaxation was adequate for the surgery. Blood transfusion was required during the procedure. The patient was alert within three minutes after discontinuation of the nitrous oxide. The patient did well postoperatively and neurological examination of the lower extremities was unchanged from the preoperative examination. The child was discharged from the hospital on the fifth postoperative day.

DISCUSSION

Little is known of the way pediatric patients with early onset myotonic dystrophy respond to anesthesia. The anesthetic management of the present patient was influenced by considerations in the anesthetic care of patients with myotonic dystrophy (adult form) which include impairment of skeletal, cardiac and smooth muscle function with multisystem involvement. A caudal anesthetic was chosen because it is a safe, reliable and simple way to produce surgical analgesia in infants and children.⁴ We anticipated that the block would lead to a quiet comfortable recovery with a shorter period of stay in the recovery room and provide some postoperative analgesia.

The use of halothane for induction and caudal anesthesia for maintenance was considered preferable to continuing the halothane for maintenance without the caudal. Our aim was to induce the child with the minimum concentration of halothane needed to permit insertion of needle into caudal canal. The use of halothane alone would have required higher inspired concentrations and prolonged administration of the agent in order to intubate the trachea and provide adequate operating conditions. This would have made the patient more prone to postoperative shivering which has been associated with the development of myotonic crisis.⁵ In addition, the contin-

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Received from the Department of Anesthesiology, The University of North Carolina Chapel Hill, Chapel Hill, North Carolina 27514. Accepted for publication May 19, 1981.

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Key Words: Anesthetic techniques: epidural; caudal. Anesthesia: pediatric. Complications: myotonic dystrophy.

ued use of halothane may have led to disorders of cardiac conduction and cardiac decompensation as has been reported during enflurane anesthesia in a patient with myotonic dystrophy.⁶

Before the final selection of a caudal as the anesthetic technique of choice, we also considered the potential risks of this procedure in this patient which included hypotension, nausea, vomiting, systemic toxic reactions to local anesthetic, high or total spinal block, and partial or complete failure to achieve an adequate block subsequently requiring supplementary anesthetic. However, when we weighed these risks against those resulting from prolonged general anesthesia, our feeling was that most of the problems with caudal anesthesia were easily preventable and manageable.

The routine use of general anesthesia for correction of skeletal deformities may be unnecessary in a majority of cases with early onset myotonic dystrophy. It is undesirable to anesthetize the entire patient, whose disease involves the muscles of respiration for an area of relatively limited surgery. Spiegel⁷ believes that caudal anesthesia is the regional anesthesia of choice for surgery for

the abdomen, perineum and lower extremities in patients two years of age and younger. Since the myotonic syndromes involves muscles themselves and not their innervation, conduction anesthesia may not produce adequate relaxation. For this reason the use of a caudal anesthetic in pediatric patients with myotonic dystrophy should be limited to lower extremity surgery not requiring excessive muscle relaxation.

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Obstruction of an Endotracheal Tube by Lidocaine Jelly

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Lidocaine jelly is frequently used as a lubricant for endotracheal intubation.¹ We describe a case in which an endotracheal tube was obstructed with this lubricant during laryngeal surgery. We also describe the results of a comparative study of lidocaine jelly and ointment.

REPORT OF A CASE

A 50-year-old woman was scheduled for microsurgical resection of a laryngeal polyp. Otherwise, preoperative examination did not reveal any significant abnormal findings. Her past history was unremarkable. One hour after administration of 0.5 mg atropine and 75 mg hydroxyzine, im, anesthesia was induced with 400 mg thiamylal and 60 mg succinylcholine, iv. A wire-reinforced anode endotracheal tube with an inflatable cuff was then inserted using lidocaine jelly as the lubricant.

After confirming that the tube was in the trachea, the cuff was inflated. Anesthesia was maintained with halothane and nitrous oxide. Surgery was performed uneventfully with the patient supine and the head extended and supported by a suspension laryngoscope. After surgery, the head was returned to the normal position. Soon after suctioning from the endotracheal tube, breath sounds became feeble by auscultation of both lung fields. Both the inspiratory and expiratory phases of ventilation were prolonged. Deflation of the endotracheal tube cuff did not alleviate the apparent airway obstruction. The endotracheal tube was then removed and replaced with a 7.0 high-volume, low-pressure endotracheal tube. The symptoms of upper respiratory tract obstruction disappeared. The patient was monitored for one hour after which the trachea was extubated with no further respiratory problems.

METHODS

Lidocaine jelly, 2 ml, was applied to five anode and five-low pressure, high volume endotracheal tubes. Lidocaine ointment, 2 ml, was also applied to five anode and five low-pressure, high-volume endotracheal tubes. All tubes were exposed to a flow of 60 per cent nitrous oxide and oxygen (4 liter/min).

RESULTS

After one hour of exposure to this gas flow, all the tubes coated with the lidocaine jelly formed the same

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Key words: Airway: obstruction. Equipment: tubes, endotracheal. Anesthetics, local: lidocaine jelly.