ANAESTHESIA FOR ACHONDROPLASTIC DWARFS UNDERGOING CAESAREAN SECTION

F. CARLI (*), E. H. THOMAS (**)
(*) M.D., F.F.A.R.C.S., Anaesthetic Registrar,
(**) M.B.B.S., D. Obst. RCOG., F.F.A.R.C.S.,

SUMMARY

The special problems related to the anaesthetic management of achondroplastic patients undergoing Caesarean section are discussed. A brief review of the recent literature and case reports of two patients who underwent Caesarean section are presented.

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INTRODUCTION

Caesarean section is the elective method of delivery in achondroplastic dwarfs as a result of the disproportion between a normal sized baby and a contracted and narrow pelvis ([11]). Problems related to the altered physiology may occur during pregnancy and during anaesthesia for Cesarean section.

Two cases of achondroplastic women who underwent General Anaesthesia for Cesarean section are reported and the anaesthetic implications discussed.

CASE REPORT

1A. D., 29 year old, an achondroplastic dwarf woman was admitted to the hospital for elective Caesarean section in July 1978. A previous Caesarean section performed in 1976 under general anaesthetic was uneventful. An achondroplastic female baby was delivered with an Apgar score of 10, five minutes after delivery. The baby died 5 weeks later and cot death was notified.

Physical examination revealed a 132 cm, 66 kg woman with short limbs, large cranium, prominent lumbar lordosis and pronounced frontal bossing. After the second trimester she developed supine hypotensive syndrome. The patient was positioned on the operating table with a wedge under the right pelvis.

Anaesthesia was induced with Thiopentone 250 mg and Suxamethonium 75 mg. Cricoid pressure was applied and endotracheal intubation and ventilation were performed. A 3130 kg female achondroplastic infant with poor muscle tone was delivered. Apgar score at 5 min. after delivery was 5. Peri- and postoperative course of both mother and child were uneventful.

2. C. R., 31 year old woman, an achondroplastic dwarf, was admitted to the hospital for elective Caesarean section in December 1979. She had received several general anaesthetics in the past for orthopaedic surgery to the legs. Two previous Caesarean sections in 1974 and 1977 under general anaesthesia were uneventful.

The patients weight was 51 kg and height 120 cm. She had a thoracic kyphosis and lumbar lordosis. Being aware of the problems presented by regional anaesthesia, a general anaesthetic was given. The anaesthetic was uneventful and a normal male infant was delivered without problems.

DISCUSSION

Achondroplasia is transmitted as an autosomal dominant trait with a high mutation rate and with an incidence of 17-29 per million in the United Kingdom ([6]). The pathology consists of a defect of endochondral ossification producing underdevelopment and premature ossification of bones formed from cartilage ([8]). Patients affected by this malformation rarely exceed 140 cm in height ([5]).

Achondroplastic patients present the following features which may be of importance to the anaesthetist:

1) There is protrusion of the forehead, depression of the root of the nose, a short and broadened maxilla and a pronounced mandible.

2) There is a defect of cartilage proliferation at the base of the skull with resulting reduction in size of the foramen magnum.

3) The spinal canal is restricted throughout its length, leading to a diminution of the subarachnoid and extradural spaces ([12]).

4) Thoracolumbar kyphosis is usually present.

5) The sacrum and the pelvic inlet are narrowed ([11]).

In achondroplastic women the combination of kyphoscoliosis, accentuated lordosis and narrow pelvis displaces the uterus anteriorly and superiorly towards the thoracic cavity. This displacement causes a more pronounced decrease in functional residual capacity (FRC) than in normal patients resulting in severe impairment of cardiorespiratory function and a tendency to hypoxia after the 28th week of pregnancy. Hypotension resulting from compression of the great vessels by the enlarged uterus in a limited abdominal cavity is more likely to occur in achondroplastic patients during pregnancy and delivery. Mrs. A.D. suffered from supine hypotensive syndrome in the third trimester of her pregnancy.

General anaesthesia has been recommended as the technique of choice for Caesarean section for achondroplastic dwarfs ([13]). Adequate lateral tilt, fluid preload and preoxygenation are necessary to avoid hypotension and hypoxia before induction of general anaesthesia. During induction some difficulty may arise with maintenance of the airway owing to the facial features, and limited extension of the atlanto-occipital joint. Extension or flexion of the neck may be impaired making intubation difficult ([14]). Hyperextension of the neck should also be avoided because acute cord compression can be precipitated ([4]). As these patients may undergo several operations for orthopaedic or neurological disorders, access to patients' notes for any previous anaesthetic problems may be helpful. Precautions should be taken such as preoperative X-ray of the cervical spine in any dwarf who has a history of neurological symptoms of spinal cord or nerve root compression. The cervical spine is often involved and odontoid dysplasia associated with atlanto-axial instability has been reported ([13]). In this situation neck flexion presents severe risk of cord compression and, in one case reported, the head was kept hyperextended throughout the entire course of anaesthesia.

Regional anaesthesia may be considered in a patient who wishes to remain awake during Caesarean section. This method however should be carefully evaluated because of the difficulties of the technique in these patients and the problems associated with it ([1]). Owing to the relatively short and narrow spinal canal containing a normal sized spinal cord, there is a high incidence of spinal tap, difficulty in assessing the dose of local analgesic and its spread ([1]). The same Author claimed the advantages of extradural over subarachnoid block for the former allows titration of dose requirements with anaesthetic block level. Nevertheless both blocks,

when they are attempted, present risk of endangering the spinal cord, because 46% of these patients have a history of spinal complications ([2, 6]). The neurological complications following spinal stenosis occur most commonly after the third or fourth decade of life, but, because severe dorsolumbar kyphosis may be present before this age group, symptoms of spinal cord or nerve root compression can occur earlier ([3, 9]).

Spinal stenosis can cause neonatal death as a result of a shortened skull base and a small foramen magnum ([10]). The same Author reported that 80% of affected infants die during the first year of life due to narrowness of the foramen magnum. This seems the most likely cause of death of A.D.'s baby (see above) who died suddenly at 5 weeks.

In conclusion, general anaesthesia remains the technique of choice for many Authors. Regional anaesthesia may be considered in a patient who wishes to remain awake but the possible technical difficulties should be explained to the patient and she should be prepared for a general anaesthetic if local anaesthesia is unsuccessful.

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