ANAESTHETIC MANAGEMENT OF ACHONDROPLASIA

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Achondroplasia is the commonest cause of the short-limbed type of dwarfism. It is inherited as an autosomal dominant gene: the great majority of cases are the result of mutation. Females are affected more frequently than males. Since it is characterized by retardation of endochondrial bone formation the basic abnormality is a disturbance of cartilage formation, mainly at the epiphyseal growth plates and at the base of skull (Fairbank, 1978). Mental and sexual functions are normal (Eilert, 1980). Muscular development is normal. Prematurity is frequent, but those surviving the first year of life generally have a normal life expectancy (Fairbank, 1978). The predicted height for achondroplastic males is 132 cm and for females is 122 cm (Nehme, Riseborough and Tredwell, 1976). Hypochondroplastic individuals are normally taller than those with achondroplasia. Hypochondroplasia usually does not involve the skull, other features being similar to those of achondroplasia. Orthopaedic sequelae, such as kyphoscoliosis and genu varum are common in achondroplastic patients (Wynne-Davies, Walsh and Gormley, 1981).

SUMMARY
We report a patient with achondroplasia who presented for emergency Caesarean section. Endotracheal intubation was not difficult. The reaction of this patient to drugs such as thiopentone and tubocurarine, judged on body weight basis, was normal.

CASE REPORT
A 34-year-old Nigerian female, a known achondroplastic dwarf, was admitted to the University of Benin Teaching Hospital, Benin City, Nigeria, in March 1984. At the time of the anaesthetic consultation she was 38 weeks pregnant by date. She had two live babies, and no abortions. The first child, male, was born in 1975 at the General Hospital, Sapele, near Benin City, by an “operation”. The second child, male, was born in 1980, again by an “operation” at the U.B.T.H., Benin City. Unfortunately, the details of these “operations” and the anaesthetic management on both of these occasions could not be traced. However, the patient recalls that she was “put to sleep” on both occasions. The operations were uneventful.

Present history
Her last menstrual period was on 27.6.83 and the expected date of delivery was 4.4.84. The course of pregnancy was uneventful. She was scheduled to undergo an elective Caesarean section on 22.3.84; no prior anaesthetic consultation was sought. However, the operation was not performed and she went into labour in the early morning of 26.3.84. An emergency Caesarean section was planned.

Physical examination
Clinically, the patient had the typical look of an achondroplastic dwarf. Her height was 98 cm, and weight 32 kg. Her cranial vault was quite large and the bridge of nose remarkably depressed. Her arms and thighs were very short. The trunk looked quite large. She had marked kyphoscoliosis. She showed marked anxiety about her short stature, pregnancy and the impending operation. In addition, she appeared to be in a state of respiratory embarrassment and pain. Her heart rate was 82 beat min⁻¹ and regular. Arterial pressure was 110/60 mm Hg in the sitting position. Heart sounds were found to be normal on auscultation. The trachea was centrally placed. The chest movements were
symmetrical, but the expansion was much less than that in other Nigerian females of her age. The air entry was diminished in both the lungs. Fine basal crepitations could be heard bilaterally. The reports of measurements of haemoglobin concentration and PCV, or urinalysis were unavailable. There was no other abnormality.

**Anaesthetic management**

Premedication consisted of atropine 0.5 mg i.v. Following pre-oxygenation for 3 min, anaesthesia was induced with 2.5% thiopentone; the patient required 120 mg. Suxamethonium 40 mg was used to facilitate the insertion of a 6.5-mm cuffed endotracheal tube. Intubation was not difficult although, on laryngoscopy, her epiglottis appeared to be high and anteriorly placed. Sellick's manoeuvre was used to safeguard against possible regurgitation.

Anaesthesia was maintained with nitrous oxide 4 litre : oxygen 3 litre in a semi-closed circuit with a soda lime absorber. Tubocurarine was used to provide adequate muscle relaxation and intermittent positive pressure ventilation (IPPV) was established. IPPV was carried out manually. It required more effort than normal to ventilate the lungs. Arterial pressure decreased from 100 mm Hg systolic to 60 mm Hg following the tubocurarine, but responded well to an increase in the rate of infusion of 5% Dextrose solution. Arterial pressure remained around 80 mm Hg (systolic) throughout the operation. Surgery was difficult: the incision to delivery time was long (25 min). A female baby weighing 2.45 kg with Apgar scores of 7/1, and 10/5, was born. The baby looked abnormal. The entire operation lasted for 150 min. The total tubocurarine administered during the 150 min was 20 mg. Reversal of residual neuromuscular blockade was achieved satisfactorily with neostigmine 2.5 mg and atropine 1.2 mg. Spontaneous ventilation was quickly established. After extubation of the trachea, the patient was given oxygen by face mask for 10 min in the operation theatre.

The blood loss was estimated to be about 500 ml and required replacement; plasma expanders were transfused as blood was not available. Blood-stained urine was obtained on catheterization, and continuous bladder drainage was instituted for 14 days after operation.

On 6.4.84 the patient's respiratory volumes were studied: her FEV$_1$ was 67.3% and FVC was 0.52 litre. The patient was followed up for 2 months.

**DISCUSSION**

The literature on the anaesthetic management of achondroplastic patients is scanty. Although Mather (1966; cited in Atkinson, Rushman and Lee, 1976) warned of the difficulties with tracheal intubation as a result of abnormalities at the base of the skull, endotracheal intubation in this patient on the present occasion was not difficult. Presumably, there was no difficulty on two previous occasions also. One of us (G.N.K.) anaesthetized a 22-year-old Iraqi female achondroplastic patient for an elective Caesarean section in 1978 (unpublished data) and did not find any difficulty in endotracheal intubation. Rimoin (1979) stated that there is more clinical and radiographic variation in achondroplastic patients than is generally described and that, as a result, every patient with achondroplasia does not necessarily pose difficulty in endotracheal intubation.

The requirement for thiopentone for the induction of anaesthesia, as judged on a body weight basis, was normal, although it was much less than the expected dose in a normal Nigerian female of her age. Similarly, the requirement for tubocurarine on a body weight basis was normal in this patient, but again much less than the normal adult female dose. This suggests that drugs should be administered on a precise body weight basis in achondroplastic patients. Their responses appear normal.

Kyphoscoliosis has been described as a frequent orthopaedic sequela in achondroplasia (Wynne-Davies, Walsh and Gormley, 1981). This patient had a marked kyphoscoliosis. Increased efforts were needed to maintain adequate ventilation manually, as a result of the structural and functional changes in the thoracic cage and lungs (Spencer, 1978). It is suggested that pre-oxygenation before anaesthesia and the administration of oxygen by face mask for some time after endotracheal extubation may be beneficial. Because of the diminished respiratory function, postoperative management should include adequate physiotherapy to avoid any possible postoperative respiratory complications.

Regional analgesic techniques may be technically difficult because of the lumbar lordosis, and the narrowing of the spinal canal. In elderly achondroplastics, the possibility of disc herniation with
resultant paraplegia (Fairbank, 1978) may militate against the use of subarachnoid blockade as the technique of choice. In young achondroplastics in whom lumbar lordosis is not marked, subarachnoid blockade may be possible. In 1978, one of us (G.N.K.) performed a successful subarachnoid blockade on a 25-year-old Iraqi male achondroplastic patient who presented for the open reduction of fractures of both bones of the right leg. More experience of patients with achondroplasia is necessary before one could comment upon the usefulness of extradural or Bier's blockade.

The anxiety shown by these patients is much greater than that observed in normal adults. A good rapport between anaesthetist and patients may prove useful in elective cases.

In this patient, because of various factors such as the non-availability of x-ray films, reagents etc., even routine investigations could not be carried out. It is suggested that a radiological skeletal survey should be carried out in any patient with achondroplasia. Any deformity of the base of the skull may predict a possible difficult intubation. X-ray can also help in differentiating achondroplasia from hypochondroplasia.

REFERENCES


