ANAESTHETIC PROBLEMS ASSOCIATED WITH WEAVER’S SYNDROME

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A child with the clinical features of Weaver’s Syndrome presented for surgery twice within a period of 6 weeks. Weaver’s Syndrome is a very rare condition characterized by accelerated growth beginning prenatally, advanced osseous maturation, unusual craniofacial appearance, herniae and camptodactyly.

CASE REPORT

A 14-month-old male child weighing 34 kg, with accelerated skeletal growth and the abnormal facies of ‘Weaver’s Syndrome’ (fig. 1), was admitted to the King Edward VIII Hospital, Durban for investigation. He was found subsequently to have an incarcerated left inguinal hernia and was scheduled for an emergency laparotomy. Pre-operative assessment of the child highlighted a number of potential problems. The child’s head was the size of an adult (57 cm in circumference) (fig. 2), but the jaw was small and the chin receding (fig. 3). The larynx lay anterior and cephalad. The child’s mouth was of normal size for his head, but his teeth were underdeveloped and his tongue was large and thickened. The child’s neck was short and broad. Consequently his ability to extend his neck was limited. He had a marked kyphosis with an angle of flexion of 65°. He had excessive amounts of very mobile subcutaneous fat, both around his neck and on his limbs. No veins could be seen or palpated.

Fortunately, the hernia was reduced manually before the child received his premedication, and it was decided to postpone surgery and repair the hernia as an elective procedure.

Next day, the child received ketamine 2 mg kg⁻¹

SUMMARY

A child of 14 months with the clinical features of Weaver’s Syndrome presented for surgery twice within a period of 6 weeks. The prime anaesthetic difficulty associated with this case, namely a difficult airway, and other problems related to Weaver’s syndrome, are discussed.
i.m. in theatre to facilitate a gaseous induction of anaesthesia. This produced sufficient sedation to allow the insertion of an i.v. cannula. After gaseous induction with halothane and nitrous oxide in oxygen, direct laryngoscopy was performed using a large MacIntosh blade. The epiglottis could not be seen and visualization of either vocal cords or laryngeal inlet was impossible. Anaesthesia was continued with a Guedel airway (No. 3) and a mask (No. 3) without difficulty. The airway was easy to maintain and the child breathed nitrous oxide, oxygen and halothane spontaneously via a Magill (Mapleson A) system. The patient made an uneventful recovery.

He presented a second time, 6 weeks later for repair of a substantial umbilical hernia. Premedication was with diazepam syrup orally 2 h before operation, but this proved inadequate and ketamine 2 mg $\text{kg}^{-1}$ i.m. was given.

The same anaesthetic technique was used for this procedure as for the previous operation on the inguinal hernia and the child's airway was never compromised. A further attempt was made to visualize the vocal cords, but only the tip of his rigid V-shaped epiglottis could be seen.
DISCUSSION

Weaver's Syndrome is a rare overgrowth syndrome beginning prenatally with accelerated skeletal maturation, unusual facies and camptodactyly. First described by Weaver and colleagues (1974) there have been a number of reported cases since (Moreno et al., 1974; Gemme et al., 1980; Majewski et al., 1981; Weisswichert, Knapp and Willich, 1981), but the patient described has the grossest features of any of the previous cases presented in the literature. Of the seven cases reported by the above authors and ourselves, six had herniae—either inguinal or umbilical and in some cases, both. There is a significant risk, therefore, of small bowel obstruction in these children and, therefore, a need to protect the airway from aspiration during emergency surgery.

We were fortunate in this child not to require tracheal intubation to protect the airway. Obviously, had emergency surgery been undertaken such a need would have arisen, to prevent the aspiration of stomach contents.

It is worthy of note that the child first presented for anaesthesia at the age of 1 month, when a general anaesthetic was given for a scan of his enlarged head. At that time the anaesthetist had great difficulty in locating the larynx, but ultimately succeeded in passing a size 4 mm uncuffed oral tube through an apparently normal larynx. At that age, the patient's head was smaller and presumably the laryngeal anatomy not as distorted as on the later occasions described here.

White and Kander (1975) suggested that the most important factor determining the ease of direct laryngoscopy was the posterior depth of the mandible. When large, this was thought to hinder displacement of soft tissues. Other important measurements reported were: an increase in the anterior depth of the mandible and a decrease in the distance from the occiput and the spinal process of C1. Cass, James and Lines (1956) considered the most important factors to be: short muscular neck and a full set of teeth; receding lower jaw; protruding upper incisors and poor mobility of the temporo-mandibular joint. However, a recent study by Van der Linde, Roelofse and Steenkamp (1983) compared eight jaw measurements and five spinal measurements from 13 patients in whom difficulty had been experienced during direct laryngoscopy, with the same measurements in 13 control patients. They used xerographic radiographs for their measurements and could find no statistical difference between the two groups. They concluded that no single factor determined the ease of direct laryngoscopy, but that a combination was important.

Our patient had a series of skull radiographs performed to assess his anatomy, as he is now being considered for spinal surgical correction of his gibbus before this defect causes any neurological damage. As this child had poorly developed teeth and full jaw movement, these factors presented no problems. However, his bull neck and receding lower jaw caused difficulties because of the position of his larynx.

A number of methods have been described to facilitate tracheal intubation in difficult patients. Awake intubation (Duncan, 1977), with either local anaesthetic spray or a superior laryngeal nerve block, is a well documented and safe method of managing patients with abnormal laryngeal anatomy, but only applicable to patients who are co-operative, which was not the case with the patient presented here.

The technique of blind nasal intubation in the anaesthetized patient as advocated by Defalque (1971) could have been utilized, but it was considered that this would have subjected the child to unnecessary naso-laryngeal trauma in a patient in whom a tracheal tube was not essential. Bearman (1962) described the use of a hook, and Waters (1963) suggested that a catheter passed through the cricothyroid membrane could be used as a guide for the tracheal tube. Murphy (1967) supported the use of a fibreoptic endoscope and Taylor and Towey (1972) a fibreoptic bronchoscopy as aids to intubation. While the fibreoptic equipment is helpful in expert hands, it is not a suitable alternative for an anaesthetist with little or no prior experience in its use. One is also limited by the size of the scope, which prevents the use of the smaller tracheal tubes. Cricothyroid puncture may well have aided intubation, but perhaps the least invasive method of protecting the child's airway would have been to have used an oesophageal obturator as described by Don Michael, Lambert and Mehran (1974) and Merrifield and King (1981).

Weaver's Syndrome presents a challenge to the anaesthetist, especially when the clinical manifestations are as pronounced as they were in the reported case. The major problem appears to be the position of the larynx which, although normal for a child of 14 months, was inappropriately placed for his adult-sized head. Should he present...
again for corrective surgery of his gibbus, intubation may be effected by the blind nasal route or with a fibreoptic device, once the dimensions of the laryngeal inlet have been determined. These could perhaps be ascertained using computerized axial tomography.

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REFERENCES


