DIFFICULT INTUBATION: A HAZARD IN THALASSAEMIA
A Case Report

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SUMMARY
A case is described which illustrates how hypertrophy of the maxillary bone marrow in thalassaemia may complicate endotracheal intubation.

Thalassaemia [thalass (Greek) = sea], or Mediterranean anaemia, is a type of haemolytic anaemia which is known to occur among people of widely differing ethnic origins (Went and MacIver, 1961). It is an inherited defect involving the haematopoietic and osseous systems. Two factors are involved, heredity and the extent of inheritance. The severe form of the disease (thalassaemia major) is the expression of homozygous inheritance whereas the mild or symptomless types (thalassaemia minor and thalassaemia minima) denote a heterozygous make-up (Whitby and Britton, 1963). There is a disturbance of haemoglobin metabolism. This results in the appearance of abnormal red blood cells having a high content of foetal haemoglobin F, which replaces some or all of the normal haemoglobin A, depending upon the degree of severity of the disorder.

Shapiro and Poe (1955), Browne (1965) and Gilbertson (1965) have described the anaesthetic management of patients with gross haemolytic anaemia, particularly in sickle-cell disease, where a genetic abnormality results in the presence within the red cells of haemoglobin S. In thalassaemia sickling of the red cells does not occur but the same principles of management apply.

CASE REPORT
A happy and alert Negro girl aged 11 years, was admitted to hospital with a mucocele of the left lacrymal sac. She was suffering from thalassaemia. This condition was affecting all her bones and she had a most unusual leonine facial deformity. Anaemia, jaundice, hepatomegaly and splenomegaly were obvious clinically. Examination of the chest revealed no pathology and her cardiovascular system appeared to be within normal limits.

Investigations.
Haemoglobin electrophoresis showed 71.5 per cent foetal Hb and a raised Hb A, of 2.5 per cent. Frequent haemoglobin estimations varied between 8 and 9 g/100 ml. Other values were as follows: p.c.v. 30 per cent, m.c.h.c. 28.4 per cent, r.b.c. 3,200,000/cu.mm., reticulocytes 3.8 per cent, target cells +++; spherocytes +, serum iron 157 nig/100 ml. The VDRL flocculation test and the Reiter complement fixation test for syphilis were non-reactive. Chest radiography showed clear lung fields. A skeletal survey showed marked hypertrophy of the bone marrow.

She was scheduled for dacryocystectomy and premedicated with atropine 0.6 mg. Following pre-oxygenation a sleep dose of thiopentone was given, and this was followed by suxamethonium 50 mg. Using an adult bledaded Macintosh laryngoscope, intubation was attempted. Although the muscles were fully relaxed and the mandible was freely mobile and normal in appearance it was not possible to view the glottis directly. The position of the head and neck were carefully adjusted and an assistant applied pressure to the thyroid cartilage. This maneuver made it possible to see the posterior surfaces of the arytenoid cartilages. The massive forward protrusion of the maxillae, prevented a normal view of the glottis and only the back of the pharynx could be seen (figs. 1 and 2). An oro-pharyngeal airway was placed in position and the lungs

FIG. 1
Forward protrusion of the maxillae, preventing direct view of the glottis.

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were ventilated with a mixture of oxygen, nitrous oxide and halothane. The distal end of a malleable stilette was curved sharply and inserted in an endotracheal tube so that the lower end of the latter could be placed in the trachea, using the posterior aspect of the arytenoid cartilages as a guide. This was done atraumatically but not without some difficulty. The operation continued uneventfully and the postoperative course was satisfactory.

DISCUSSION

The investigations in this case placed the patient’s anaemia midway between a major and a clinically affected minor thalassaemia. Family haemoglobin studies carried out subsequently showed the father’s blood picture to be quite normal, and the mother to have a thalassaemia trait. This child was therefore suffering from thalassaemia minor, despite her high level of foetal haemoglobin and extensive bone changes. Patients with thalassaemia major usually die before puberty from intercurrent infection or from liver and cardiac insufficiency caused by haemochromatosis. The clinical picture in the less severe forms of the illness is that of a haemolytic anaemia. The condition has many of the features of sickle-cell anaemia. The bone changes that occur resemble the hypertrophic marrow changes found in sickle-cell anaemia but in some respects are more exaggerated. Considerable overgrowth of the maxillae may develop which impedes pneumatization of the paranasal sinuses (Middlemiss, 1961).

Our experience in this case indicates that in addition to the general problems of managing cases of this kind, the possibility of a difficult intubation should be borne in mind.

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REFERENCES


INTUBATION DIFFICILE—UN RISQUE DE LA THALASSÉMIE: RAPPORT D’UN CAS

Sommaire

L’auteur décrit un cas qui illustre comment l’hypertrophie du moelle osseuse maxillaire en cas de thalassémie peut compliquer l’intubation endotracheale.

SCHWIERIGE INTUBATION EINE GEFAHR BEI DER THALASSÄMIE: EIN FALLBERICHT

Zusammenfassung

Es wird ein Fall beschrieben der zeigt, wie eine Hypertrophie des Oberkieferknochenmarks bei der Thalassämie die endotracheale Intubation komplizieren kann.