pending upon the severity of the operation. This unit is equipped with new adjustable beds, oxygen tent, piped oxygen and suction apparatus to each bed.

CONCLUSIONS

With the increasing demands made by ill patients on hospital facilities, it is important that we adopt the newer and more economical progressive patient care and principles. This method of treatment and care is being adopted by more and more hospitals in America and Europe, and we, in this country, should give this matter serious attention.

SUMMARY

The need for progressive patient care is stressed. Care in hospital can be divided into intensive care, intermediate care and self care. These various units are described. A suggestion is also made as to the role of the general practitioner in this scheme.

REFERENCES


PENDRED'S SYNDROME IN SOUTH AFRICAN BANTU BROTHERS

SOLOMON E. LEVIN, M.B., M.R.C.P. (EDIN.), D.C.H. AND LEON H. KLUGMAN, M.B., M.R.C.P., Department of Paediatrics, Baragwanath Hospital and Department of Medicine, General Hospital and the University of the Witwatersrand, Johannesburg

In 1896, Vaughan Pendred reported the association of goitre and deaf-mutism in 2 female siblings of an Irish family. Brain described 5 families with 12 cases of simple goitre associated with congenital deaf-mutism; he suggested a recessive mode of inheritance for this condition. It was not until 1958 that the metabolic defect in thyroxine synthesis was identified when Morgans and Trotter found that, following the administration of a dose of $^{131}I$, a proportion of the trapped $^{131}I$ was discharged from the thyroid gland by the subsequent administration of potassium perchlorate. This finding suggested a partial defect in the organic binding of iodine. Despite this enzyme defect, most of the patients are euthyroid. Fraser et al. studied 18 families with 28 affected subjects and showed the deafness to be high-tone perceptive in type, the mode of inheritance simple recessive in nature and that $^{131}I$ in the thyroid was partly discharged by potassium perchlorate in all their cases. Recently Fraser has presented a study of 207 families. He mentions that Pendred's syndrome has been reported among persons originating from the British Isles, Germany, Belgium, Holland, France, Italy, Greece, Sweden, Denmark, Poland, the Lebanon, the USA and Canada. Three Indian children described by Fraser represent the first report of this syndrome in non-White people.

We therefore thought it of interest to record the occurrence of Pendred's syndrome in 2 brothers, who are South African Bantu subjects.

CASE REPORTS

Case 1

T.M., a Bantu male aged 7 years, was admitted because of a swelling in the neck of a week's duration; he had been deaf and dumb since birth. His appetite was good and he had no pain or pressure symptoms that could be ascribed to the goitre. He did not come from an endemic goitre area.

Examination showed a well-nourished child with a large diffuse goitre, the right lobe being larger than the left. There was a systolic bruit over the gland. The heart rate was 80/min. and the blood pressure was 110/80 mm.Hg. There was no clinical evidence of hyper- or hypothyroidism. The height was 51 in. (129.5 cm.), which was at the 90th percentile, and the weight was 64 lb. (29 kg.), also at the 90th percentile. The following investigations were carried out: Mantoux test—negative; mumps complement-fixation test—negative on 2
which increased on ward diet, as shown in Table I. During this time, it was noted that the goitre had diminished in size, though it remained quite large. Measurement of the neck circumference ranged between 12 and 12½ in.

Treatment with 1-thyroxine sodium was commenced at the beginning of January 1964, and the dose was increased to 0.1 mg. twice daily by March 1964. The goitre steadily decreased in size and a neck measurement in May 1965 was 10½ in.

Family history. The father was 73 years old and in good health. Two children were born of his first marriage, both males, now aged 25 and 21 years respectively; they are both normal.

Case 2

L.M. was a Bantu male aged 11 years, weighing 76 lb. (34.5 kg.), and with a height of 56 in. (148 cm.), both measurements being at the 50th percentile. He was deaf and dumb and his nutritional status was satisfactory. The pulse rate was 72/min. and the blood pressure was 120/80 mm.Hg. There was a diffuse enlargement of the thyroid gland with the right lobe more prominent than the left. The over-all size of the goitre was less than that of his brother. There were no clinical signs of hyper- or hypothyroidism.

The following investigations were done: Mantoux test was negative; X-ray of the chest was within normal limits; the bone age was 10-12 years as indicated by X-ray films of the elbow. The haemoglobin was 13.7 G/lOO ml. The leucocyte count was 6,000/cu.mm., and the platelets were normal. The serum proteins were 7.8 G/lOO ml. of which 4.3 G was albumin. The protein-bound iodine was 6.3 mcg./100 ml and the blood cholesterol 190 mg./100 ml.

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Audiometry demonstrated a bilateral perceptive high-tone deafness, the left ear being more severely affected than the right. The 24-hour 131I-uptake was 34%. Subsequently the 131I-uptake at 1 hr. was found to be 18.3% and 30 min. after the administration of potassium perchlorate the radioactivity over the thyroid had fallen to 5% of the 1-hr. value (Fig. 1). A scintigram showed diffuse uptake over the thyroid gland.

Treatment commenced with 1-thyroxine sodium in a dose of 0.3 mg. daily. The neck circumference was 12½ in. on admission and 12½ in. 1 year after the commencement of therapy.

TABLE I. PBI AND CHOLESTEROL LEVELS IN THE BLOOD OF CASE 1

<table>
<thead>
<tr>
<th>Date</th>
<th>PBI µg./100 ml</th>
<th>Cholesterol mg./100 ml</th>
</tr>
</thead>
<tbody>
<tr>
<td>4/10/1963</td>
<td>3-8</td>
<td>190</td>
</tr>
<tr>
<td>25/10/1963</td>
<td>6-8</td>
<td>190</td>
</tr>
<tr>
<td>12/12/1963</td>
<td>6-0</td>
<td>185</td>
</tr>
<tr>
<td>9/3/1964</td>
<td>9-5</td>
<td>190</td>
</tr>
</tbody>
</table>

DISCUSSION

These cases fulfil the criteria for the diagnosis of Pendred's syndrome, namely, the triad of goitre, high-tone perceptive deafness and a partial discharge of radio-iodine from the thyroid gland after the administration of potassium perchlorate. Both patients were euthyroid and this has been the case with the vast majority of reported cases. However, a few children have presented in early life as goitrous cretins. More commonly, the goitre appears in middle childhood and even later in the case of males.

As a result of the partial block in thyroxine synthesis, it is thought that low blood levels of thyroxine stimulate the production of thyroid-stimulating hormone (TSH) by the anterior pituitary, thus inducing thyroid hyperplasia and the production of a goitre. This constant stimulation results in an extremely pleomorphic histological change in the thyroid gland, which has been interpreted by some authors as evidence of malignancy. However, Fraser is of the opinion that malignant change probably does not occur in these cases.

The use of thyroxine in treatment is aimed at suppression of the excessive TSH production in the hope of reducing the size of the goitre. A striking response was obtained in the younger of our 2 patients, but not in his older brother.

The recessive mode of inheritance of this condition is supported by the family history obtained. The first wife with 2 normal children was presumably not a carrier of the necessary gene defect, while the second wife and the father must be heterozygous carriers for 2 instances of Pendred's syndrome to have appeared among their 4 children.

Pendred's syndrome is not of infrequent occurrence in institutions for the deaf. Thould and Scowen found an incidence of approximately 2% among children examined at 15 such schools. Fraser found an incidence of 5.6% at normal and specialized schools for the deaf. This condition is, therefore, a numerically important cause of congenital deafness.

Deaf-mutism together with other forms of defective physical and mental development are known to occur with endemic goitre; in these cases, however, there is an absence of thyroidal discharge after potassium perchlorate. Pendred's syndrome has also been reported from an endemic goitre area in the Lebanon.

REFERENCES


SUMMARY

Two Bantu brothers with the classical features of Pendred's syndrome are reported from South Africa. These are thought to be the first such cases described from the African continent.

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