HYPOTHYROIDISM FOLLOWING THYROIDECTOMY FOR CONGENITAL OBSTRUCTIVE GOITER

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THE first report of congenital goiter was written by Fodéré in 1792.[3] Since that time many cases have been described in the European and Scandinavian literature.[1,4] In contrast to this, very few instances have been recorded in American publications.[2, 6, 9, 13, 15, 17] The first case of congenital goiter in Ohio or Minnesota, where the incidence of simple goiter is high, was not reported until 1939. In all the cases so far reported in the United States the mother showed some evidence of thyroid dysfunction, and in most of the cases had been treated with or received one of the iodine compounds as expectorants either before or during pregnancy. One report failed to indicate whether the mother did or did not receive iodine.[6] In the case to be described, the mother at no time received any of the iodine compounds. The child was born with an obstructive goiter which was removed surgically. Symptoms of hypothyroid function followed operation, and these were rectified by replacement therapy.

CASE REPORT

On Nov. 4, 1948, B. V., a 5-week-old male infant, was admitted to The Mount Sinai Hospital. The patient was the result of the third pregnancy; the two siblings were normal children, 16 and 4 years of age. This child was born at home after a full-term pregnancy and normal delivery. At birth, three lumps were noted in the infant's neck. The birth weight was unknown. During the first week the infant experienced difficulty in swallowing, with several choking episodes. Peculiar gurgling sounds in the throat were heard. During the ensuing four-week period the sounds had increased in severity and had been associated with tachypnea. The size of the cervical masses had not been observed to have increased materially. The infant had never become cyanotic, and it was noted that he preferred to lie with his head hyperextended.

The parents were both natives of New Jersey. There was no family history of any thyroid disturbance. The paternal grandparents were born in Holland and settled in the United States in 1890. Of possible significance is the observation that the mother had had several "nervous breakdowns" in the past, and at the time of the infant's admission was suffering from her most recent attack. She had never taken any thyroid medication or iodine-containing compounds.

On admission the child was a well-developed and well-nourished infant with a good cry, not hoarse, having no respiratory difficulty other than noisy respirations. The weight was 8 pounds, 9 ounces. The length was 21 inches. The head was maintained in hyperextension and could not be flexed without producing dyspnea. In the neck there were three masses. The submandibllar space and anterior triangles of the neck on either side were filled with soft, somewhat cystic, fairly well-demarcated masses. The mass on the right extended posteriorly beyond the angle of the jaw, and inferiorly into the base of the neck. The mass on the left was smaller and the inferior margin extended just to the base of the neck. The masses did not transilluminate, and both extended

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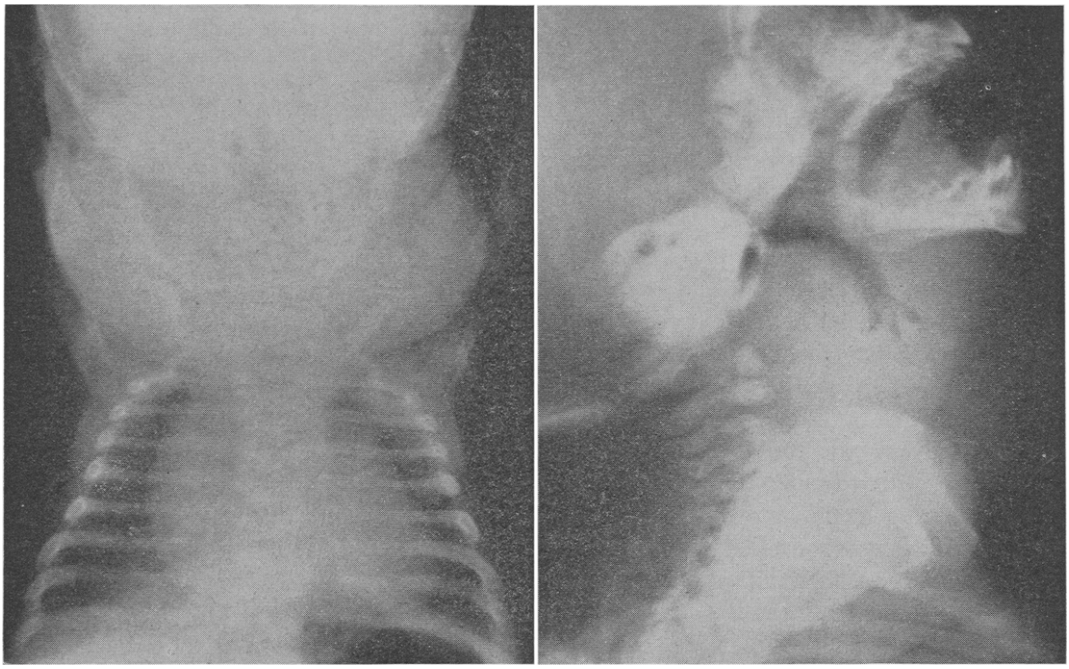
toward the midline where there was a third mass almost golf ball in size. No definite distinction could be made between the masses. In the midline, all the masses were below the level of the hyoid bone. The bridge of the nose was low. The cheeks were chubby and the eyelids were slightly puffy. The mouth was kept open, but there was no macroglossia and the tongue did not protrude. Digital palpation of the hypopharynx revealed a mass anteriorly by the masses described. No intrathoracic component was present. (Fig. 1, *A* and *B*.) The differential diagnosis on admission included cystic hygroma of the neck and multiple cysts of the thyroid.

On November 18, the patient was operated upon under intratracheal anesthesia by one of the authors (E. S. H.), with Dr. Ernest Arnheim. A curvilinear collar incision was made extending completely across the base



A. *B.*

Fig. 1.—*A*, Preoperative roentgenogram, anteroposterior projection. The rounded masses bulge from both sides of the neck. *B*, Preoperative roentgenogram, lateral projection. The head is hyperextended, and the hypopharynx markedly displaced anteriorly.

in the retropharyngeal area which was pushing the trachea and pharynx anteriorly. The other positive finding on admission was a grade 3 systolic murmur heard best at the base of the heart. No thrills were palpable and there was no precordial bulge. A small umbilical hernia was present.

The hemogram and the urine examination were normal. Roentgenograms of the neck revealed the esophagus and trachea to be displaced of the neck, with division of the platysma and strap muscles. It was apparent that the pathology was confined to the thyroid gland, which grossly had the appearance of a large goiter. All of the lobes were greatly enlarged, and the left lobe extended around and behind the larynx. The larynx was not completely encircled. The gland was uniformly firm in consistency and there was greatly increased vascularity. Because of considerable doubt as to the nature of

the underlying disease, a frozen section was performed on an excised portion of the gland. The tentative diagnosis was neoplasm of the thyroid and total thyroidectomy was advised. The left superior pole vessels were isolated, doubly ligated, and divided. The isthmus was mobilized and divided to the left of the midline. A total excision of the left lobe was performed with division of the middle and inferior vessels on the capsule of the gland. The right lobe was similarly mobilized and resected together with the isthmus, leaving only a small segment of thyroid tissue 1 cm. in diameter at the right inferior pole. At the right lower pole there was a firm marble-sized nodule which was removed together with the remainder of the lobe.

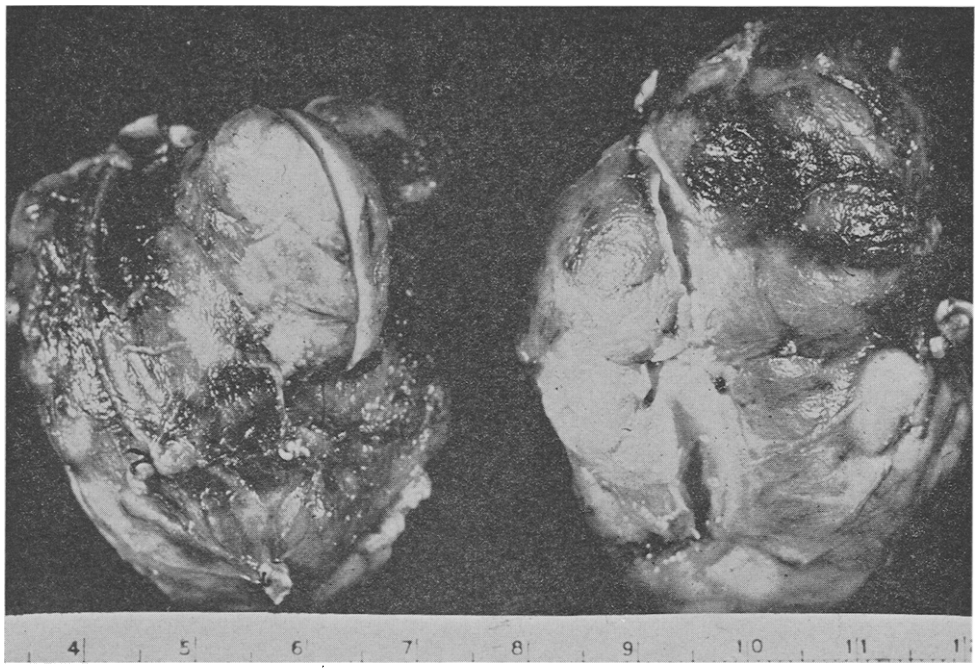
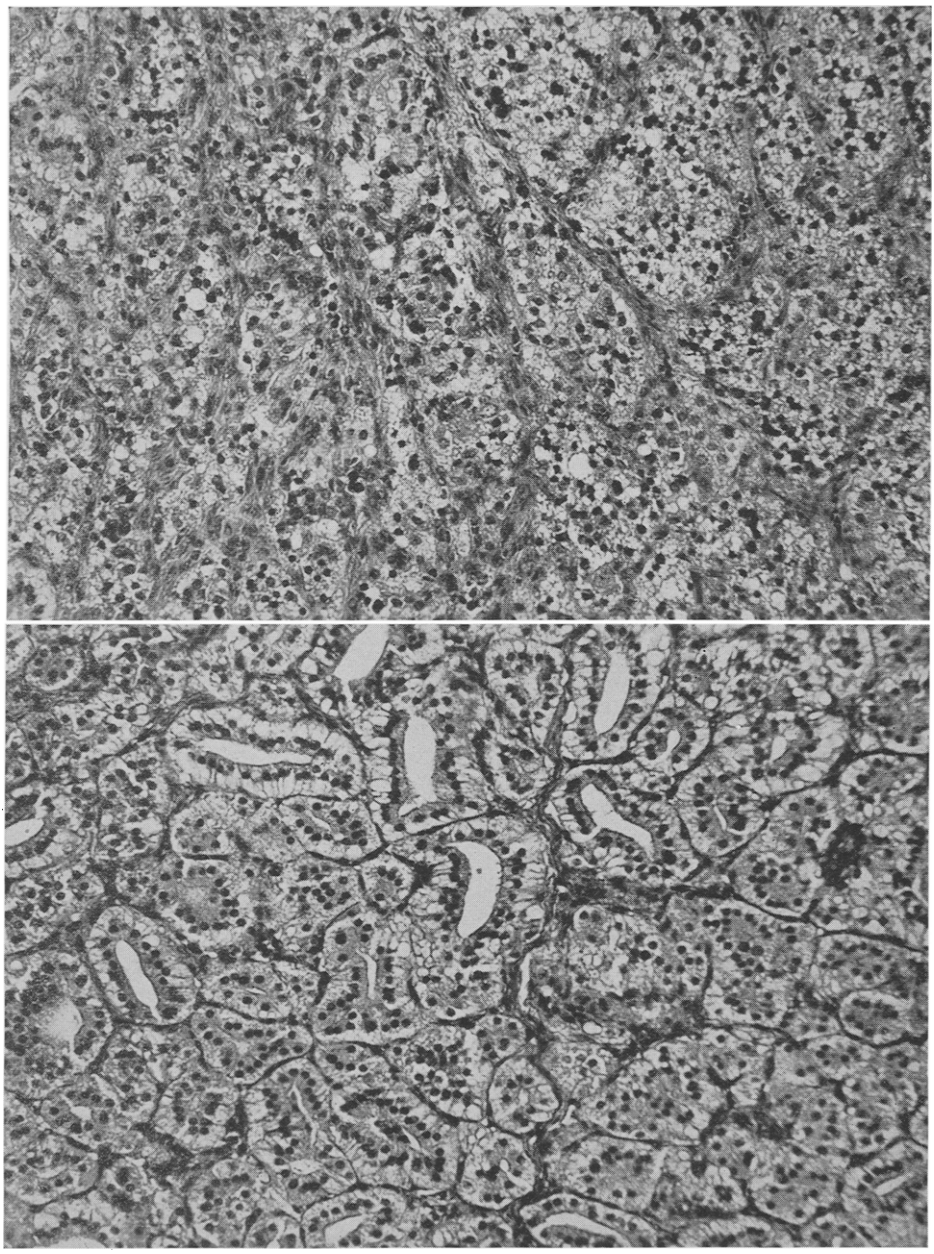


Fig. 2.—Anterior view of both resected lobes of the thyroid gland.

The gross description of the mass and the microscopic examination revealed the following: The specimen consisted of an elongated semilunar mass of thyroid tissue weighing a total of 65 grams. The mass measured 13 × 5 × 2.5-3.0 cm. (Fig. 2). The entire mass was enclosed in a thin fibrous transparent capsule. A dilated segment of vein coursed in a transverse direction along the anterior surface of the specimen. Upon opening the vein it was found to contain a moderate amount of clotted blood but appeared to be free of neoplastic invasion. Section of the mass revealed a soft, gray pink, cellular tissue irregularly arranged in variable-sized lobulations by very fine fibrous trabeculae. The microscopic report was thyroid showing changes compatible with a diagnosis of congenital diffuse parenchymatous stroma. (Fig. 3, *A* and *B*.)

The infant's postoperative course was uneventful except for moderate laryngeal edema which was secondary to the intubation anesthesia and the surgical manipulation in the laryngeal area. This was successfully treated by steam tent, frequent suctioning, and Adrenalin in the form of an aerosol. Tetany was not detected at any time.

A.



B.

Fig. 3.—Histologic structure of thyroid gland. Hematoxylin and eosin stain, ×185. *A*, The disorganized architecture in this area suggested a malignant lesion on frozen section. *B*, Typical acinar architecture, characteristic of most of the resected gland.

Laryngoscopy revealed normal motion of both vocal cords. The head could now be flexed normally, respiratory obstruction was eliminated, and there were no further episodes of choking during feeding. A roentgenogram no longer showed the tremendous anterior displacement of the trachea (Fig. 4).

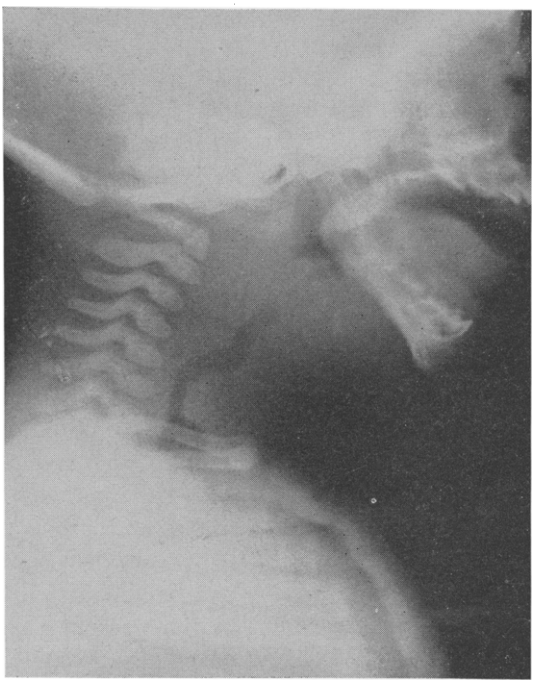


Fig. 4.—Postoperative roentgenogram, lateral projection. The head can now be flexed, and much of the anterior displacement of the airway has been corrected.

In order to determine whether the patient had been receiving adequate thyroid hormone from the goitrous gland, roentgenograms were taken for bone age two days after operation. These revealed the presence of epiphyseal centers at the lower end of the femur and upper end of the tibia. The patient's bone age was therefore commensurate with his chronological age of eight weeks. Roentgenograms of the wrists failed to reveal the presence of carpal centers. The serum cholesterol value was 280 mg. per cent.

Dessicated thyroid was started on the seventh postoperative day as replacement therapy. Thyroid, U. S. P., ¼ grain, was given daily for eight days and then increased to ⅜ grain daily for nine days. At the end of the seventeen-day course of thyroid the drug was stopped. There was no change in the infant's appearance during the period of treatment. It was the intention of the pediatrics department, in stopping the drug, to give radioactive iodine after two weeks had elapsed, to determine whether the patient had any functioning thyroid tissue left. However, it was the opinion of the radiophysics department that, should the radiation be taken up by the thyroid gland, there might be sufficient radiation to destroy whatever function of the gland still remained. The child was therefore discharged from the hospital on Dec. 17, 1948, on the twenty-ninth postoperative day, to return two weeks hence for repeat serum cholesterol determinations. The serum cholesterol on discharge was 200 mg. per cent and the alkaline

phosphatase was 18 King-Armstrong units.

In spite of attempts to follow the patient, he was not seen again until March 9, 1949, when he was readmitted to the hospital. During the three months since discharge, the patient had not received any thyroid and the family stated that the child had been well, but had difficulty in eating because his tongue seemed to be getting larger. He was constipated, and glycerin suppositories had been given frequently. The weight was 13 pounds, 6 ounces, and the length was 23½ inches. In three months the child had gained 4 pounds in weight and had grown 2½ inches in length. The facies had changed somewhat. The child manifested a thick, protruding tongue, which was grossly and uniformly enlarged, puffy eyes, low-bridged nose, and a general apathetic appearance. The clinical impression at this time was hypothyroidism secondary to almost complete thyroidectomy. The serum cholesterol was 240 mg. per cent three and one-half months after the discontinuation of thyroid. This was a rise of only 40 mg. per cent over the level at the time of discharge. The serum alkaline phosphatase was somewhat low for this age, being 7 to 11 King-Armstrong units. Roentgenograms of the lower end of the femur and upper end of the tibia revealed the epiphyseal centers formerly present. The plasma bound iodine level was lower than 4 gamma, which is strongly suggestive of hypothyroidism.

Accordingly, on March 19, 1949, thyroid replacement therapy was reinstituted. One-half grain daily was given for five days; the dose was then increased to ¾ grain for six days, and then to 1 grain daily. On this regimen the child definitely improved. He took his formula much more avidly, he appeared much brighter, his tongue became smaller and his cheeks were less puffy, and his bowel habits improved. He had one stool daily without the necessity of suppositories or enemas. The serum cholesterol level dropped to 160 mg. per cent. The patient was discharged on April 10, 1949, the thirty-second hospital day.

The patient was last examined at 27 months of age. He was still taking 1 grain of thyroid daily. His mental and physical status were commensurate with his chronological age.

DISCUSSION

The preoperative diagnosis of extensive cystic hygroma of the neck was based in part on the clinical appearance of the lesion; the fact that congenital goiter is so rare in this region practically excluded it from consideration. Hygromas need not transilluminate, particularly if hemorrhage has occurred within the cysts. They may be a cause of mechanical distress. While usually unilateral, bilateral involvement has been seen, as well as involvement of the mediastinum and other parts of the body.[4] However, the firm consistency and anatomical distribution of the masses in the present case should have suggested the correct diagnosis.

In this case the diagnosis of congenital goiter was made after practically total thyroidectomy had been performed. Following surgery it was our intention to maintain the patient on thyroid replacement therapy. However, the parents did not follow the program outlined upon discharge of the patient, and no thyroid had been given for three and one-half months, when the patient was brought back to the follow-up clinic. By this time definite signs of hypothyroidism had supervened. The tongue was coarse and thick and protruded from the gaping mouth. The hair was sparse; the umbilical hernia was more marked. In addition, constipation

and failure to suck well were present. The clinical impression of hypothyroidism was not substantiated by the initial laboratory results. The roentgenograms revealed normal bone development for the patient's age, and two epiphyseal centers in the wrist had developed while the patient was not receiving thyroid replacement therapy. The serum cholesterol had not increased appreciably while treatment had been withheld. According to Wilkins[18] when thyroid medication is discontinued in hypothyroid children the serum cholesterol increases greatly in the course of four to twenty weeks, reaching levels 98 to 410 mg. per cent above those of the treated period. In normal children the rise is only 10 to 55 mg. per cent. The serum alkaline phosphatase was only moderately low for a 5½-month-old child, being 7 to 11 King-Armstrong units. The only chemical determination which was strongly suggestive of the diagnosis of hypothyroidism was the plasma protein bound iodine value of less than 4 gamma. Salter and associates state that a plasma protein bound iodine level of less than 4 gamma per liter is definitely subnormal and is of distinct value in confirming the diagnosis of poor thyroid function.[12] Two weeks after thyroid replacement therapy had been started, definite changes in appearance and behavior were noted. The child was last seen at the age of 27 months. He appeared physically and mentally normal and was still taking thyroid medication.

When the diagnosis of congenital goiter is made prior to surgical intervention, the application of iodine-containing ointments to the skin over the gland, plus the oral administration of iodine compounds, have been recommended. It is claimed that the goiter will frequently disappear after this treatment. On the other hand, Rienhoff has described rapid relief of dyspnea and cyanosis in a newborn infant with a very large thyroid, and grossly detectable shrinkage in the size of the gland after a few days of therapy with thyroxin administered hypodermically; in his experience iodine has been ineffectual.[11]

The most striking symptom in patients with congenital goiter other than the mere presence of the mass in the neck is the respiratory difficulty secondary to pressure on and displacement of the trachea. In the majority of the cases reported in the American literature respiratory obstruction was so severe that suffocation and death resulted. In cases where the respiratory symptoms are severe enough to produce dyspnea and cyanosis, emergency life-saving surgery must often be performed. Transection of the isthmus of the thyroid may afford relief from the mechanical obstruction in some of these cases. Because of retrolaryngeal extension together with compression in the tracheo-esophageal grooves, partial and subtotal thyroidectomy have often been required. It is felt that tracheotomy is less desirable in this neonatal age group than either of the other procedures described.

A summary of cases of congenital goiter treated surgically is contained in Table I. In each case respiratory obstruction required relief. Seven infants survived thyroid resection, in contrast to two newborn infants succumbing after tracheotomy. In one instance (D'Abreu[1]), left hemithy-

TABLE I. CASES OF CONGENITAL OBSTRUCTIVE GOITER TREATED SURGICALLY

CASE	AUTHOR	LOCALE	MATERNAL THYROID DISEASE	AGE AT OPERATION	SURGICAL PROCEDURE	SIZE OF RESECTED GLAND	PATHOLOGIC REPORT	RESULT
1	Peterson and Sondern,[10] 1911	New York City	Not stated	5 weeks	Subtotal resection	Horseshoe 6 × 4 × 3 cm. and 4 × 2.5 × 2 cm.	Adenoma and colloid degeneration	Good
2	Williamson,[19] 1933	New Orleans	None in 3 generations	18 months	Subtotal resection	25 grams	Diffuse adenomatous hyperplasia	Good
3	Symmers,[16] 1941	New York City	Not stated	6 weeks	Subtotal (4/5) resection	47 grams	Hürthle-cell tumor	Good
4	Davies,[2] 1943	Vancouver	Adenomatous goiter (untreated)	13 days (3rd goitrous baby)	Subtotal resection	6 grams	Subinvoluted phase of hyperplastic gland	Good
5	Morrow,[8] 1945	Chicago	Not stated	2 months	Part of each lobe plus isthmus	4 × 2 × 1.5 cm.	Hürthle-cell tumor	Good
6	Kunstadter,[6] 1948	Great Lakes	Not stated	Newborn	Tracheotomy	(Autopsy) 62 grams	Struma hyperplastica	Died
7	Seligman and Pescovitz,[13] 1950	New York City	Hyperthyroid (treated)	Newborn	Tracheotomy	(Autopsy) 16 grams	Hyperplastic gland	Died
8	D'Abreu,[1] 1950	England	None	1.1 month 2.3 months	1. L. hemithyroidectomy 2. R. hemithyroidectomy	4 × 3 × 2.5 cm. 4.5 × 3 × 2.2 cm. (14 grams)	Hyperplastic gland c̄ incipient involution	Good
9	Authors' case, 1948	New York City	None	7 weeks	Almost total resection	65 grams	Congenital diffuse parenchymatous struma	Good c̄ thyroid replacement therapy

roidectomy resulted in only temporary improvement, necessitating right hemithyroidectomy two months later before lasting relief was obtained. No reports of a fatality following thyroid resection for congenital obstructive goiter have been found. In at least three of the cases there was no evidence of maternal thyroid disease. There was no sex predilection. The possible significance of the diagnosis "Hürthle-cell adenoma" in Cases 3 and 5 is beyond the scope of this discussion.

Had the condition been properly identified in the present instance, a less extensive resection would undoubtedly have been preferable from the point of view of subsequent glandular function. Postoperative hypothyroidism was not recorded in any of the other surgically treated patients. Estimates of the normal weight of the thyroid in the newborn infant range from 0.7 grams[13] to 1½ to 2 grams.[1] The weight of the resected gland in the present instance (65 grams) is greater than that recorded in any of the patients treated surgically, as well as the autopsied cases summarized by Jones.[5] However, huge congenital goiters, resulting in severe dystocia and marked obstetrical difficulties, have long been recognized in the European goiter belts. McLanahan[7] delivered a stillborn fetus of a goitrous primipara; the fetal goiter measured 10⅞ inches in largest diameter.

SUMMARY

1. The incidence and clinical features of sporadic congenital goiter are presented.

2. A case of congenital obstructive goiter with surgically induced hypothyroidism in a 5-week-old child is described.

3. Criteria for proper diagnosis and therapy are discussed.

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