provisional diagnosis being an irreducible right inguinal hernia. Under general anaesthesia a right oblique inguinal incision was made, dividing the right external oblique aponeurosis. This exposed the mass, which was found to be a collection of thrombosed vessels in the substance of the round ligament. This was divided, excised, and ligated at the internal abdominal ring. A small hernial sac was found to be empty; it was transfixed and excised. The inguinal canal was closed by suture of the conjoint tendon to the inguinal ligament. The post-operative recovery was uneventful, the patient leaving hospital on March 2. On April 14 healing of the right groin was perfect, and there was no evidence of a left inguinal mass.

It was thought that the "left inguinal hernia" in this case was a varicocele of the round ligament which subsided after the pregnancy, whilst a smaller varicocele of the right round ligament which was not discovered during pregnancy became thrombosed immediately after delivery, probably owing to the alteration in the size of the uterus producing a circulatory disturbance; both varicoceles were in association with varicose veins of the vulva.

Discussion

We believe that nearly all groin tumours first appearing in pregnancy are due to varicosities of the round ligament, which at first sight simulate an inguinal hernia. While in 2 out of the 12 cases under review there was little evidence of varicose veins, apart from the varicocele of the groin, in the remaining 10 varicosities of the vulva, with or without similar lesions of the thigh, were present, often to a gross degree. In seven patients a diagnosis of inguinal hernia was made when the mass was first seen, before being referred to us, and in one case operation had been advised.

It does not appear likely that a hernia of the inguinal region should make its first appearance during pregnancy, for, although there is an increase in intra-abdominal pressure, most of the structures which would fill the hernial sac (with the exception of free fluid) are kept out of the way by the bulk of the pregnant uterus and broad ligament. Some authorities (Berkeley *et al.*, 1938; Baird, 1950) state that pre-existing herniae frequently disappear during the later months of pregnancy, "the reason being that the enlarging uterus pushes the intestines away from the inguinal ring and presently blocks access to them" (Berkeley *et al.*, 1938). Strangulated inguinal herniae are extremely rare in the later months, and, in fact, Gaudier (1894) states that a strangulated inguinal hernia and pregnancy are incompatible. Although it is generally agreed that known pre-existing herniae tend to be temporarily cured during pregnancy, this series of 12 cases of swellings in the inguinal region first appearing in pregnancy was collected in the short space of 12 weeks. It is known that early in pregnancy, in the first trimester, there is a great increase in the vascularity of the pelvis, and varicose veins of the lower extremity may appear or worsen before the size of the uterus is likely to affect the venous return. The veins of the round ligament form an anastomosis with those of the vulva, and will join in any generalized varicosity—thus a varicocele of the round ligament is formed.

Clinical Features.—The clinical features of varicoceles of the round ligament are similar to those of an inguinal hernia. The patient complains of a lump and an aching feeling in the groin. The situation is identical, since both veins and hernial sac accompany the round ligament in the inguinal canal. Both swellings disappear on lying down and reappear on standing, though a varicocele may return more gradually than a hernia and is a little less circumscribed. A varicocele is of course dull on percussion and more closely resembles a hernia containing omentum than one containing bowel. The signs and symptoms of both conditions are therefore very similar, and it is only possible to make a diagnosis by the fact that in the case of the varicocele the swelling appears for the first time during pregnancy. The absence of varicose veins elsewhere does not exclude the diagnosis of a varicocele of the round

ligament. Thrombosis of the varicose veins is a complication which still further increases the resemblance between an inguinal hernia and varicocele of the round ligament, and is described in Case 2. This complication produces a firm, tender, irreducible swelling simulating a strangulated inguinal hernia, and a differential diagnosis becomes an urgent necessity, in view of the entirely different treatments required.

Treatment.—No active therapy has been found necessary in the patients we have seen. Suggestions to operate during pregnancy indicate that the probable aetiology of the lesion has not been appreciated. It is not necessary to commit a patient to bed rest because of the condition, although it is known that this occurred in a patient elsewhere, not in our series, in the belief that an inguinal hernia was present. A truss may, by giving support, add to the comfort of the patient. Reassurance, with a simple explanation of the temporary nature of the lump during pregnancy, and the recommendation of the usual supportive measures for the relief of vulval and leg varicose veins, is all that has been found necessary.

We wish to express our thanks to members of the clinical staff of Queen Charlotte's Maternity Hospital for helping to collect the cases, and to Mr. Bailey, of Queen Charlotte's and Chelsea Hospital photographic department, for the photographs.

REFERENCES

- Baird, D. (1950). *Combined Textbook of Obstetrics and Gynaecology*, 5th ed. Livingstone, Edinburgh.
 Berkeley, C., Bonney, V., and MacLeod, D. H. (1938). *The Abnormal in Obstetrics*. Arnold, London.
 Gaudier (1894). *Rev. Chir. (Paris)*, 14, 859.
 Verovitz, C. H. (1941). *Diseases of the Veins and Lymphatics of the Lower Extremity*. Christopher Publishing House, Boston, U.S.A.

THE SYMMETRICAL FORM OF STATUS BONNEVIE-ULLRICH (TURNER'S SYNDROME)

BY

J. B. BARLOW, M.B., B.Ch.

AND

S. E. LEVIN, M.B., B.Ch.

Department of Paediatrics, Baragwanath Hospital,
Johannesburg

In 1930 Ullrich reported a case of multiple congenital anomalies in an 8-year-old girl that was typical of a syndrome since known on the Continent as the symmetrical form of status Bonnevie-Ullrich. This child was of short stature, had a webbed neck (pterygium colli), an increased carrying angle of the arms (cubitus valgus), hypoplastic nipples, a highly arched palate, drooping angles of the mouth, hyperflexibility of the shoulder-joints, and irregular short nails of the hands and feet. The upper eyelids were prominent and heavy, giving the impression of bilateral ptosis. The ears were closely attached to the side of the head. At birth there had been a marked pitting oedema of the nuchal region and the extremities. The oedema, "congenital lymphangiectatic oedema," disappeared during the second year of life.

Ullrich (1949) observed that this syndrome is identical with Turner's syndrome and the syndrome of ovarian agenesis, which have since been described and discussed in the English language literature by authors evidently unaware of his original report. It is apparent, however, that, whereas Ullrich (1930) described the congenital anomalies in great detail, it was the endocrinological aspects which aroused most interest in the American

and British literature. It is our purpose to present a typical case of this syndrome occurring in an African infant.

Case Report

A 3-weeks-old Bantu female was first seen on April 8, 1953. The mother complained that the infant had been "swollen" since birth.

Physical Examination.—**Head:** The circumference of the head was 14 in. (35 cm.). There was marked pitting oedema of the scalp, especially over the occipital region. Some oedema was present round the eyes, and the epicanthic folds were prominent. The bridge of the nose was flattened and somewhat broad. The corners of the mouth were drooping and the palate was highly arched. The upper parts of the ears were bound to the margin of the scalp. The superficial scalp veins were unusually conspicuous and large. The cranial nerves appeared normal. **Neck:** There were broad loose folds of skin over the postero-lateral aspects of the neck (pterygium colli). The occipital hair-line was low. **Chest:** The skin over the whole of the back was loose (cutis laxa). The chest was somewhat broad in shape. The nipples were hypoplastic, being pin-point in size. The heart and lungs were clinically and radiologically normal. **Abdomen:** The tone of the abdominal musculature was normal. A small umbilical hernia was present. **Extremities:** There

Photograph showing pitting oedema of the extremities.

was marked pitting oedema of the extremities (see Fig.). The nails of the fourth and fifth toes of the left foot and the fifth toe of the right foot were absent. The remainder of the nails of the hands and feet were spoon-shaped. There was a laxity of all the joints of the limbs, and the elbow-joints could be passively placed into a marked degree of cubitus valgus. **Genitalia:** The external genitalia appeared normal.

Investigations.—The urine was normal. Haemoglobin, 13 g. per 100 ml.; blood urea, 10 mg. per 100 ml.; serum proteins, 6.8 g. per 100 ml. (albumin 3 g., globulin 3.8 g.); thymol turbidity test, 1 unit; thymol flocculation, Takata-Ara, and colloidal red tests were negative; serum sodium, 350 mg. per 100 ml. X-ray plates of the spine, skull, and extremities showed no skeletal abnormalities. The mother's Wassermann reaction was negative.

Examination of Family.—The parents and the two other siblings were examined clinically and radiologically. The upper parts of the ears of the father and both children were closely attached to the scalp margin, but no other abnormality was observed.

Progress.—When the child had been in hospital for one month she developed gastro-enteritis, which lasted for three days. During this period the oedema disappeared completely and recurred only when the diarrhoea and vomiting had stopped. She was discharged six weeks after this. When seen again, at the age of 6 months, the child was in

good health and mental development appeared to be normal. The circumorbital and nuchal oedema had disappeared, but pitting oedema of the extremities was still prominent.

Discussion

Turner (1938), unaware of Ullrich's report, described a triad characterized by webbing of the neck, cubitus valgus, and infantilism which he had observed in seven adult females. Numerous similar cases have since been reported by others (Sharpey-Schafer, 1941; Albright *et al.*, 1942; Varney *et al.*, 1942; Wilkins and Fleischmann, 1944; Lisser *et al.*, 1947; Del Castillo *et al.*, 1947; Dorff *et al.*, 1947; Jackson, 1950; Jackson and Sougin-Mibashan, 1953). Congenital anomalies observed by these authors, in addition to the webbed neck, cubitus valgus, and short stature emphasized by Turner, included the low hair-line at the nape of the neck, dystrophies of the ears, "shield" chest, arched palate, scoliosis of the spine, and coarctation of the aorta. Cases in a few females under the age of puberty were also recognized (Goldberg *et al.*, 1947; Silver, 1951; Cutler and Silver, 1953; Jackson and Sougin-Mibashan, 1953; Vulliamy, 1953).

Some authors have apparently avoided the term "Turner's syndrome"—firstly, because Turner (1938) considered that the sexual infantilism was of pituitary rather than gonadal origin, and, secondly, because congenital webbed neck or cubitus valgus was not invariably present. However, the term "syndrome of ovarian agenesis" is even more unsatisfactory, as the condition has been observed, though less commonly, in males (Flavell, 1943; Dorff *et al.*, 1948; McCullagh, 1948; Greenblatt and Nieburgs, 1948; Reforzo-Membrives *et al.*, 1949; Sohval, 1951; Cunningham and Harley, 1951; James, 1952; Sougin-Mibashan and Jackson, 1953; Mackenzie, 1953). It is possible that the term "Bonnievie-Ullrich-Turner syndrome," as suggested by James (1952), would do most to clarify the present somewhat disjointed situation and combine the contributions to the subject from both the Continental and the English language literature.

Our patient has shown virtually all the salient features of the syndrome as listed by Ullrich (1949). This author emphasized the frequency with which the "congenital lymphangiectatic oedema" occurs, and considered the formation of this subcutaneous fluid during prenatal life to be the main aetiological factor in the various congenital anomalies seen. In over a hundred patients reported in the English language literature, however, we have found reference to oedema in only two case reports. Silver (1951) stated that the feet of his patient, aged 2 years and 8 months, were described as having been "swollen" at birth; and Vulliamy (1953), in his report of an 8-year-old girl, described "unexplained oedema of hands and feet at birth, persisting in the feet for the first three years." Pitting oedema was a very prominent feature in our patient, and was, in fact, the reason that the mother sought medical advice. It is difficult to understand how, if it were present, such a conspicuous abnormality could have been so frequently overlooked. Ullrich (1949), however, stated that the swelling disappears during the first two years of life. As our patient is the first under the age of 2 years whose case is described in the English literature, this may well explain why oedema has so seldom been mentioned.

No definite hereditary factor has yet been discovered. Unfortunately we were able to trace only our patient's immediate relatives, but were impressed by the complete attachment of the upper parts of the ears to the scalp margin in the father and the two other siblings. This particular anomaly was seen by Sharpey-Schafer (1941) in his patient; it is apparent to a moderate degree in the photograph of the male patient of Sougin-Mibashan and Jackson (1953), and is well marked in the male African reported by Mackenzie (1953). Whether this ear anomaly represents a very mild form of the syndrome in the relatives of our patient it is at present impossible to determine, but it was considered distinct enough to merit being put on record.

ADDENDUM.—Since this paper was written a case of this syndrome associated with hypertrophic pyloric stenosis has been recognized in a 2-months-old infant (Keay and Lewis, 1954). These authors, however, described the oedema of the extremities in their patient as non-pitting in character.

We are indebted to Dr. J. D. Allen, superintendent of the Baragwanath Hospital, for permission to publish this report. We wish to thank Dr. E. Kahn, under whom the patient was admitted, for his encouragement and advice.

REFERENCES

- Albright, F., Smith, P. H., and Fraser, R. (1942). *Amer. J. med. Sci.*, 204, 625.
 Cunningham, G. C., and Harley, J. F. (1951). *J. Pediatr.*, 38, 738.
 Cutler, C. H., and Silver, H. K. (1953). *Ibid.*, 42, 352.
 Del Castillo, E. B., De La Balze, F. A., and Argonz, J. (1947). *J. clin. Endocr.*, 7, 385.
 Dorff, G. B., Appelman, D. H., and Liveson, A. (1947). *Ibid.*, 7, 807.
 — (1948). *Arch. Pediatr.*, 65, 555.
 Flavell, G. (1943). *Brit. J. Surg.*, 31, 150.
 Goldberg, M. B., Maxwell, A. F., and Smith, P. M. (1947). *J. clin. Endocr.*, 7, 11.
 Greenblatt, R. B., and Nieburgs, H. E. (1948). *Ibid.*, 8, 993.
 Jackson, H. (1950). *S. Afr. med. J.*, 24, 423.
 Jackson, W. P. U., and Sougin-Mibashan, R. (1953). *British Medical Journal*, 2, 368.
 James, T. (1952). *Edinb. med. J.*, 59, 344.
 Keay, A. J., and Lewis, I. C. (1954). *Arch. dis. Childh.*, 29, 424.
 Lissner, H., Curtis, L. E., Escamilla, R. F., and Goldberg, M. B. (1947). *J. clin. Endocr.*, 7, 665.
 McCullagh, E. P. (1948). Quoted by Sougin-Mibashan and Jackson (1953).
 Mackenzie, D. F. (1953). *British Medical Journal*, 2, 1273.
 Reforzo-Membrives, J., Trabucco, A., and Escardo, F. (1949). *J. clin. Endocr.*, 9, 1333.
 Sharpey-Schafer, E. P. (1941). *Lancet*, 2, 559.
 Silver, H. K. (1951). *Pediatrics*, 8, 368.
 Sohval, A. R. (1951). Quoted by Sougin-Mibashan and Jackson (1953).
 Sougin-Mibashan, R., and Jackson, W. P. U. (1953). *British Medical Journal*, 2, 371.
 Turner, H. H. (1938). *Endocrinology*, 23, 566.
 Ullrich, O. (1930). *Z. Kinderheilk.*, 49, 271.
 — (1949). *Amer. J. hum. Genet.*, 1, 179.
 Varney, R. F., Kenyon, A. T., and Koch, F. C. (1942). *J. clin. Endocr.*, 2, 137.
 Vulliamy, D. G. (1953). *Proc. roy. Soc. Med.*, 46, 279.
 Wilkins, L., and Fleischmann, W. (1944). *J. clin. Endocr.*, 4, 357.

Medical Memoranda

"Primary Myxoedema" in a Man of 30

Myxoedema was first described by Gull (1874) when he wrote "On a Cretenoid State Supervening in Adult Life in Women." It was Ord in 1878 who first used the term "myxoedema" to describe these cases of hypothyroidism. It is well known that myxoedema generally occurs in middle-aged and elderly women. Myxoedema in young males is uncommon, and so far as can be ascertained is either due to "pituitary myxoedema" as described by Means *et al.* (1940) or present as a true myxoedema following total thyroidectomy. It is therefore most unusual to find a case of "primary myxoedema" in a man of 30.

FIG. 1

FIG. 2

FIG. 1.—Photograph taken on October 23, 1953. FIG. 2.—Photograph taken on November 28, 1953.

CASE REPORT

A gardener aged 30 was first seen in consultation in October, 1953. He complained of a rash on the hands and forearms which was due to a plant-contact eczema. His general appearance was most striking, as shown in Fig. 1. On further questioning he admitted to the following symptoms: (1) the gradual onset during the past two years of increasing weariness, which was now extreme towards the end of the day; (2) increase in weight; (3) a preference for hot weather and a feeling of the cold "right through him"; (4) a deepening of his voice and a slowing and hesitant speech; (5) a drying of the skin—"he had not sweated for months"; and (6) his hair was becoming coarse and lifeless.

On examination there were no physical abnormalities apart from a bradycardia of 50. The serum cholesterol was 400 mg. per 100 ml. and the blood urea 67 mg. per 100 ml. of serum. Examination of the urine showed: sp. gr. 1009, sugar, nil; albumin, slight trace; no casts or red blood cells. X-ray examination of the pituitary fossa was normal.

Treatment with L-thyroxine sodium 0.1 mg. daily for a week, then 0.1 mg. twice a day, was started on October 23. Improvement was most dramatic, both in his appearance as shown in Fig. 2, taken five weeks after Fig. 1, and in the blood chemistry as shown in Fig. 3, the serum cholesterol and the blood urea returning to normal within twenty-eight days.

He felt perfectly fit and all his symptoms had disappeared. His voice was now normal, he could sweat, his hair was improving in texture, he had lost weight, and he was no longer weary.

COMMENT

This record is of particular interest as it describes a case of "primary myxoedema" in a young man which responded dramatically to a relatively small dose of thyroxine and showed a fall, not only of the serum cholesterol as would be expected, but also of the blood urea. The fall in the blood urea was presumably due to the increased metabolism. Or could it have been due to a clearing of "cholesterol-clogged" arterioles in the kidneys?

I am grateful to Dr. C. A. K. Bird, of Macclesfield Hospital, for his advice and help with the blood chemistry in this case.

F. R. M. YOUNG, M.B., B.S.,
Macclesfield, Cheshire.

REFERENCES

- Gull, W. W. (1874). *Trans. clin. Soc. Lond.*, 7, 180.
 Means, J. H., Hertz, S., and Lerman, J. (1940). *Trans. Ass. Amer. Phys.*, 55, 32.

Ventricular Tachycardia following Exercise in a Case of Angina Pectoris

It is felt that the following case is interesting enough to be placed on record.

CASE REPORT

A man aged 46 attended the out-patient department giving a history typical of angina pectoris of four months' duration. The attacks had never occurred before this, had on no occasion lasted longer than 10 minutes, and had never been severe enough to cause absence from work. Apart from "pleurisy" 10 years previously, no history of any past

FIG. 3.—Showing improvement in blood chemistry.