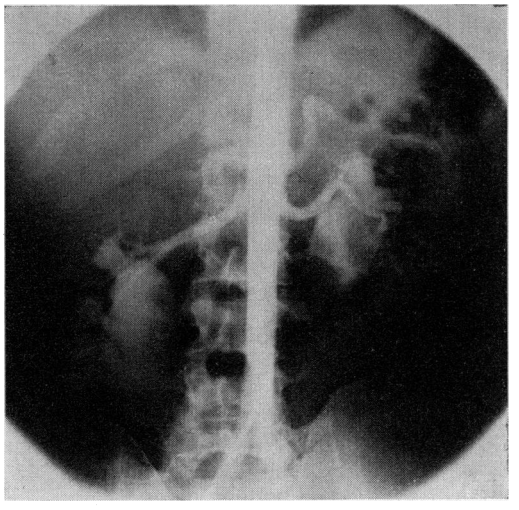

Fig 2 *Renal arteriogram showing bilateral obstruction at pelviureteric junctions and hydronephrosis*

sufficient cause for both his bilateral hydronephrosis and his raised blood pressure (Fig 2). Similarly, although a specific vascular lesion could account for the carotid occlusion, a more likely explanation is that the patient has had right cerebral atrophy since childhood, with a poor vascular supply on that side, predisposing him to vascular accidents in that area. The right carotid occlusion was probably recent, accounting for his presentation with left hemiplegia.

Neurofibromatosis is basically a process affecting schwannian elements and tissues derived from the neural crest. This is an embryologically versatile tissue and may account for the protean manifestation of the disease. It may be that a fundamental biochemical lesion affecting the neural crest produces a syndrome of which von Recklinghausen's original disease is only a part.

REFERENCES
Crowe F W, Schull W J & Neel J W
(1956) Multiple Neurofibromatosis. Springfield, Ill.
Halpern M & Currarino G
(1965) *New Engl. J. Med.* 273, 248

The President suggested that this patient had neurofibromatosis of the bladder, an exceedingly rare lesion.

The following case was also shown:
Lipoid Dermato-arthritis
Treated with Clofibrate
Dr D N Golding

Meeting February 10 1967

Case

Aneurysm of the Aortic Root with Gross Aortic Incompetence: Successful Surgical Correction

K B Saunders MRCP
and Professor H H Bentall FRCS
(*Units of Clinical Cardiology & Cardiac Surgery, Royal Postgraduate Medical School, London*)

The patient, a 32-year-old man, was referred following an insurance examination in September 1966. He had been graded A1 in the army in 1951 and 1953, and had had two further normal insurance examinations in 1959 and 1965. On direct questioning he said he had occasionally felt sensations of tightness in the chest on stooping, and had infrequently noticed a dropped beat, but was otherwise symptomless. His wife, however, had been able to hear a loud rushing noise in his chest for the previous year.

On examination there was evidence of gross aortic incompetence. He was in sinus rhythm with a collapsing pulse, and a blood pressure of 120 mmHg systolic, the diastolic pressure being unrecordable. There was moderate left ventricular hypertrophy. In the aortic area there was a soft ejection murmur, and a very loud whirring pandiastolic murmur. There was no ejection click, and the aortic second sound was not heard. No evidence of Marfan's syndrome. Electrocardiogram normal. Chest X-ray (Fig 1) was reported as showing dilatation of the ascending aorta, with unexplained prominence of the left pulmonary artery. Wassermann reaction negative. Full blood count, ESR and plasma proteins normal. Serum cholesterol 315 mg/100 ml. The presence of only moderate left ventricular hypertrophy, with a normal ECG suggested that the aortic incompetence was not of long duration, and this was confirmed by the history. A functional diagnosis of gross aortic incompetence of recent onset was made.

The anatomical diagnosis was made by aortography (Fig 2), when a large aneurysm of the root

of the aorta was demonstrated. The presence of gross aortic incompetence was confirmed.

Aneurysms at this site in the aorta are uncommon. The differential diagnosis includes syphilis, cystic medial necrosis, Marfan's syndrome, giant cell aortitis, dissection, and the Ehlers-Danlos syndrome. The prognosis of such a lesion is agreed to be extremely bad. The untreated patient faces increasing aortic incompetence and left ventricular failure, with a high added risk of sudden death from rupture of the aneurysm. Corrective surgery was therefore undertaken.

At operation the aortic valve was found to be grossly incompetent due to stretching of the valve ring, and there was a well-localized aneurysm of the ascending aorta. On the anterior wall of the aneurysm there was a very thin avascular area about one centimetre square, which might well have ruptured in the near future. The valve cusps were normal. The valve was excised; a Starr-Edwards prosthesis was then joined to a crimped Teflon prosthetic aorta and both united to the patient's valve ring. The coronary arteries were anastomosed to holes made in the wall of the graft, the latter being anastomosed to normal aorta above the aneurysm.

The immediate result of surgery is completely satisfactory. When seen three months later, the patient was symptomless and had a normal blood pressure; the valve was entirely competent.

The final pathological diagnosis was made on histological examination of the operation specimen. This showed an extremely thin aortic wall,

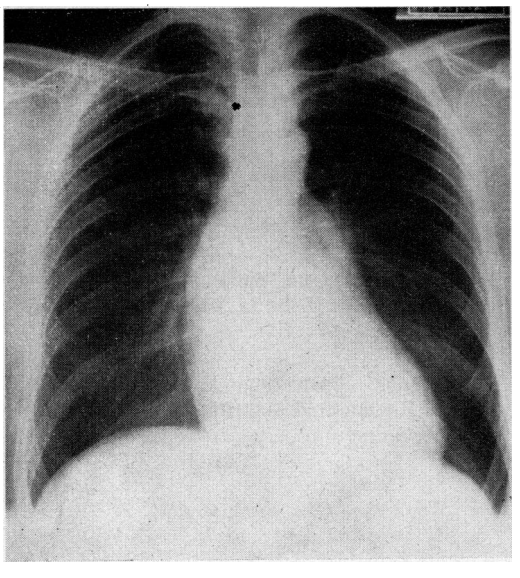

Fig 1 *Chest X-ray*

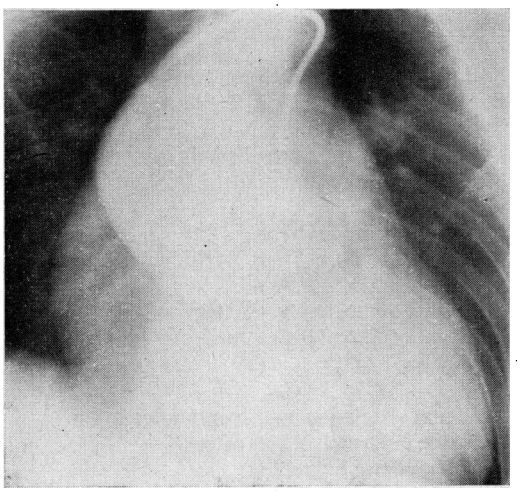

Fig 2 *Retrograde aortogram: a well-localized aneurysm of the aortic root is demonstrated. The aortic arch appears normal*

with almost complete absence of elastic tissue, appearances characteristic of Marfan's syndrome. It is stressed that there were no external stigmata of this condition.

Comment

The clinical diagnosis of such an aneurysm is difficult, for an aneurysm at the root of the aorta will rarely cause symptoms or signs by pressure on neighbouring structures, nor is its presence always obvious on chest X-ray. In retrospect, the prominence of the left pulmonary artery in this case was due to its displacement by the aneurysm: the combination of prominence of the left pulmonary artery with dilatation of the ascending aorta may prove of diagnostic value. Aortography is essential to delineate the lesion.

Although the immediate result of surgery is excellent, the final prognosis remains debatable. The first relevant factor is the fate of the valve and aortic prostheses. Such prostheses have not been in use long enough to assess the probable life which may be expected from them, although this is almost certainly more than ten years. Secondly, the patient will need anticoagulant therapy for life to prevent embolism. Finally, there is a widespread defect of elastic tissue: it may well be that further aneurysmal changes will occur in major blood vessels. Indeed, when the femoral artery was cannulated for perfusion, the wall was noted to be abnormally thin, and the vessel was dilated. This last factor may well prove to be the most important determinant of the patient's future clinical course.

Acknowledgment: We wish to thank Dr M Hamilton who originally referred the patient.

The following cases were also shown:

Islet Cell Tumour of the Pancreas
Dr A H Knight (for Dr T Parkinson)

Paraffinoma of the Large Bowel
Miss M Sheila Christian (for Mr Maurice Lee)

Tuberculosis of the Breast
Mr F A Strang (for Mr R V Fiddian)

Occlusion of the Axillary Artery in a Case of Sarcoidosis
Dr A J Karlish

Malignant Melanoma – Lipogranulomatous Reaction in Lymph Nodes after Endolymphatic Iodine 131
Mr J Blake (for Mr E Stanley Lee)

Three Cases of Ischæmic Colitis
Mr G H Dickson (for Mr Maurice Lee)

Bilateral Lower Ureteric Obstruction
Mr R R Hall (for Mr M Gough)

Meeting May 19 1967
at Wexham Park Hospital, Slough

The following cases were shown:
(1) **Megaloblastic Anæmia (Folic Acid Deficiency) due to Dietary Deficiency in an Indian Child of 2½ years.**
(2) **Portal-Hypertension due to Portal Vein Thrombosis following Exchange Transfusion, presenting with Hæmatemesis and Splenomegaly at Age of 4 years.**
(3) **15/17 Chromosome Abnormality of Patau's Syndrome**
Dr A B Donnison

Rheumatoid Arthritis: Synovectomy and Excision Arthroplasty of Metacarpophalangeal Joint Right Hand and Metacarpophalangeal Synovectomy Left Hand
Mr S Harrison and Dr B M Ansell

Rheumatoid Arthritis: McKee Arthroplasties of Both Hips
Mr G Arden and Dr B M Ansell

Rheumatoid Arthritis: Synovectomy of Right Knee, Metacarpophalangeal Excision Arthroplasty of Right Hand
Mr G Arden, Mr S Harrison and Dr B M Ansell

Calcinosis ? Secondary to Dermatomyosis
Dr B M Ansell

Dissecting Aneurysm involving Right Iliofemoral Artery
Mr E J Williams and Dr J Lister

Chlorpromazine Jaundice – Paranoid Schizophrenia and Depression
Dr P I Reed

Retroperitoneal Pseudomucinous Cystadenoma of the Ovary
Dr C B d'A Fearn (for Mr R Ramsay)

Scleroderma with Systemic Symptoms
Dr J Lister

Multiple Traumatic Urinary Fistulæ
Miss M S Christian

Cat Scratch Disease
Dr P Willcox
Boy aged 15 with gland masses in left axilla, mid forearm and left epitrochlear gland; gross suppuration was followed by surgical drainage of axillary and other abscesses; this was rapidly successful in terminating the infection, which arose from a scratch on the left hand.

The following demonstrations were given:
Demonstrations in the Pathological Department of Sugar Chromatography, Clinical Virology, Blood-gas Analysis and Cervical Cytology
Dr J A Easton and Dr F E T Scott

An X-ray Demonstration: Uncommon X-rays of General Interest
Dr E Giordani

A Film was shown on 'Suck and Drip' (the Use of the Maurice Lee Tube in Vagotomy and Pyloroplasty)

Photographs illustrating Eye Involvement in Juvenile Rheumatoid Arthritis
Mr W K Smiley