Meeting 14 March 1975

Cases

Zollinger-Ellison Syndrome

L W Lauste MD FRCS
(*Royal Sussex County Hospital, Brighton*)

Mr E G, aged 48. Office worker

The patient was presented to this Section in 1962 (Lauste 1962, *Proceedings* **55**, 806–807) and is presented again fourteen years after operation.

History: The patient, a transport driver at the time, developed indigestion in 1956 and a duodenal ulcer was diagnosed. Symptoms persisted in spite of medical treatment and in May 1960 a partial gastrectomy was performed. Persistent pain and a massive hæmatemesis led to a further gastrectomy in July 1960 for gastrojejunal ulcer. Persistent abdominal symptoms and an abdominal fistula led to a further laparotomy in November 1960. The findings were not clear.

Severe abdominal symptoms and vomiting continued and in July 1961 a subtotal gastrectomy and vagotomy for massive ulcers was performed. Two tumours were removed from the head of the pancreas: section showed islet cell adenomata.

Progress: Subsequent progress was satisfactory and he has had no further gastric symptoms.

In 1966 a severe iron deficiency anæmia developed, which responded to oral iron. In 1970 and again in 1971 the patient had severe bronchitis requiring hospital admission for investigation and treatment.

He remains well and symptom free and it seems likely that he is one of the small group in which the tumours were not malignant, and all were removed.

Severe Hypertension of Renal Ischæmic Origin

Denis G Gill[1] DCH MRCPI
(*Department of Pædiatrics, Guy's Hospital, London SE1 9RT*)

S M, boy aged 6

History: Presented in November 1973 with a six-week history of intermittent abdominal pain, anorexia and occasional vomiting.

On examination: Height 117 cm, weight 21.5 kg (both on 75th centile). Blood pressure 200/170 mmHg. Left ventricular enlargement. Grade 2 retinopathy. No murmurs and no femoral artery delay detected.

Investigations: Blood urea 50, plasma creatinine 1.1 mg/100 ml, normal plasma and urinary electrolytes, creatinine clearance 130 ml/min per 1.73 m^2, normal plasma and urinary cortisols, normal VMA excretion. Intravenous urogram was normal apart from indentations on the right ureter. The right kidney was 9 cm and the left 10 cm long. During renal arteriography the renal arteries were not visualized on free aortic injection. Selective renal arteriograms (Fig 1) showed severe stenosis of both renal arteries with post-stenotic dilatation. The right kidney received profuse collateral blood supply, particularly from the ureteric artery. Renal vein renin

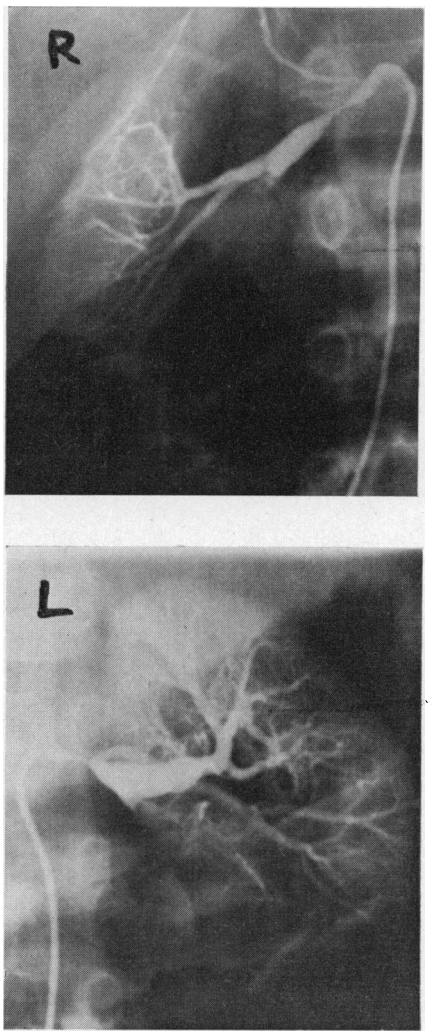

Fig 1 *Renal arteriograms*

[1]Present address: Children's Hospital, Temple Street, Dublin 1

samples were taken at the time of arteriography. All the renin values were high (Table 1) and demonstrated that the renal veins were contributing renin to the circulation. Peripheral vein angiotensin-2 was 156 pg/ml (normal 5–35). Renal scintillogram showed normally sized kidneys with good function.

The blood pressure and the drugs used in its control are shown in Fig 2. Blood pressure was labile and extremely difficult to control. The following drugs were administered simultaneously to a maximum tolerated daily dose: bethanidine 280 mg, propranolol 200 mg, diazoxide 150 mg, frusemide 240 mg, methyldopa 600 mg. Intermittent parenteral hydrallazine and diazoxide were also required.

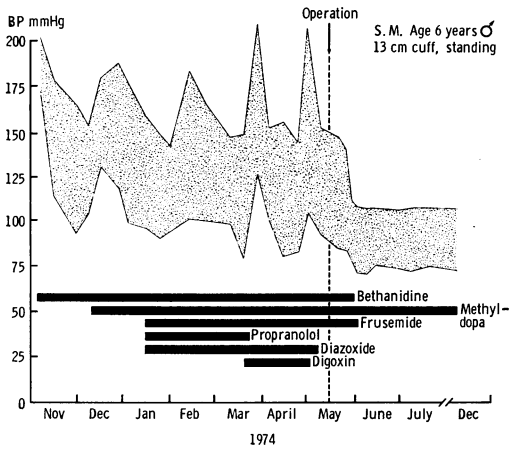

Fig 2 *Blood pressure chart showing severe hypertension, and dramatic sustained fall in blood pressure following correction of bilateral renal artery stenosis*

At operation (May 1974, Mr F Ellis): Aorta and stem vessels appeared small. Both renal arteries were hypoplastic at their origin and were almost completely occluded for a distance of 2 cm from the aorta. There was no pulsation and no recordable intra-arterial pressure distal to the narrow areas. The appearance was that of fibromuscular hyperplasia. On the left side a 6 cm graft was taken from the long saphenous vein and anastomosed between the infrarenal aorta and the left renal artery. On the right side, grafting was considered not to be feasible, and so an autotransplant was carried out, anastomosing the renal artery and vein end-to-side to the right common iliac vessels. The ureter was mobilized but left intact.

Following these procedures both kidneys revascularized fully. Urine output was satisfactory. Blood pressure promptly fell and has remained normal. Antihypertensive drugs have been withdrawn. Renal function and intravenous urogram are normal and both kidneys have grown.

Table 1

Renal vein renin studies

	Angiotensin-1 (pg/ml per hour)	*Renin concentration (units●)*
Right renal vein	26 450	463
Left renal vein	36 400	613
Inferior vena cava	13 600	288
L:R ratio	1.3	
R:IVC ratio	1.9	

● arbitrary units designated by Glasgow MRC group; normal = 4–20

Discussion

Hypertension is often unrecognized in children. Coran & Schuster (1968) reported 10 children with renovascular hypertension, of whom 7 were asymptomatic. A normal intravenous pyelogram will not exclude renal artery stenosis. Fry *et al.* (1973), in a series of 22 children with renal artery stenosis, noted that only 40% had an abnormal pyelogram. Ureteral notching was evident in one of their cases.

Measurement of renal vein renin activity and of differential renin ratios is of great value in predicting surgical cure of unilateral renal artery stenosis (Stockigt *et al.* 1972). Dillon & Reyness (1974) using a semi-microradioimmunoassay have established normal peripheral plasma renin activity in infancy and childhood.

Renovascular hypertension can be very difficult to control in children, possibly since children have much higher peripheral renin levels than adults (Godard *et al.* 1968). The results of operative treatment of unilateral renal artery stenosis in childhood are excellent, with greater than 90% cure rate (Fry *et al.* 1973). The operations of choice are by-pass grafts (Coran & Shuster 1968) and renal autotransplantation (Kaufman *et al.* 1972).

REFERENCES

Coran G C & Schuster S R (1968) *Surgery* **64**, 672

Dillon M J & Reyness J (1974) *Archives of Disease in Childhood* **49**, 823

Fry W J, Ernst C B, Stanley J C & Brink B (1973) *Archives of Surgery* **107**, 692

Godard L, Riondel A M, Veyrat R, Megevand A & Muller A F (1968) *Pediatrics* **41**, 883

Kaufman J J, Goodwin W E, Waisman J & Gyepes M T (1972) *American Journal of Surgery* **124**, 149

Stockigt J R, Collins R D, Noakes A C, Shambelan M & Bigheri E G (1972) *Lancet* i, 1194

(meeting to be continued)