Brain revascularisation in hypertension

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SUMMARY Neurological symptoms in hypertensive subjects may be a reflection of intracranial vascular disease and not just a consequence of hypertension. Two hypertensive children with renovascular disease, neurological symptoms, and severe cerebral arterial disease were treated by extracranial-intracranial arterial bypass surgery with improvement of symptoms and easier control of blood pressure. Where revascularisation surgery is appropriate, this should be undertaken before neurological complications arise.

Renal arterial disease may reflect more widespread arterial disease including that in intracranial vessels.¹ This association is not widely recognised, but is important as neurological symptoms may otherwise be attributed erroneously to the effect of the hypertension alone.

Identification of cerebral abnormalities in these patients has implications, not only because of the possibility of specific treatment for stenosed areas, as in the two children we report, but also because lowering their blood pressure may lead to cerebral ischaemia, and hence permanent neurological damage. We report two children with severe hypertension secondary to renal arterial disease, both of whom had symptoms or signs suggestive of intracranial involvement. The difficulties experienced in management and the importance of timing of surgical intervention in the treatment of the cerebral arterial abnormalities is stressed.

Case reports

Case 1. At the age of 12 months, this boy suffered a left myoclonic fit with a left hemiparesis, thought to be due to 'acute hemiplegia of infancy'; it resolved slowly. At the age of 18 months he was noted to be hypertensive, his blood pressure frequently reaching 230/170 mmHg. On clinical examination there were bilateral carotid bruits but no cutaneous stigmata of neurofibromatosis. Investigations showed a raised plasma renin activity (more than 40 000 ng A1/l per hour), normal renal function, normal urinary vanilmandelic acid excretion, and no raised porphyrins values. Renal arteriography showed multiple intrarenal arterial stenoses, and he was treated with

methyldopa, propranolol, and chlorothiazide. His blood pressure, however, remained difficult to control at around 150/100 mmHg. At age 7 years renal arteriography was repeated, and the findings suggested progressive arterial abnormalities in both kidneys. Although renal vein renin estimation showed lateralisation to the left kidney, the presence of extensive disease in both kidneys precluded operative management of his blood pressure. Because control of blood pressure remained unsatisfactory despite the addition of prazosin and minoxidil, captopril was begun at age 8 years. His hypertension was then better controlled but a few months later he woke one morning with severe headache, weakness, and pallor. He was unable to speak and his blood pressure was 120/100 mmHg. There was gradual resolution of his symptoms over approximately 24 hours. Bilateral carotid angiography showed extensive intra- and extracranial vascular occlusive disease. There was narrowing of both internal carotid arteries with bilateral occlusion at the level of the cavernous sinuses. A left extracranial-intracranial anastomosis was undertaken. His blood pressure rose initially after surgery, requiring labetalol infusion and reintroduction of captopril, but by the sixth day control was good and further recovery was unremarkable. Five months later he was admitted for right sided extracranial-intracranial anastomosis and he remained extremely well after the operation. Oral hypotensive treatment was reintroduced within two hours of completion of surgery. He made an uneventful recovery.

After three years both grafts are pulsatile, he has had no further neurological problems, and his rate of growth has been normal. His blood pressure is maintained at a level appropriate to his age (110/70

mmHg) with modest doses of captopril, propranolol, and frusemide.

Case 2. The second patient presented at 4 years of age because her mother noticed a cranial bruit. She was found to have sustained hypertension with blood pressure 180/120 mmHg and a loud intracranial bruit. Investigations showed a raised plasma renin activity but no abnormalities on renal angiography or localisation on selective renal vein renin sampling. A computed tomogram of the brain was normal, cerebral angiography detected narrowing at both carotid syphons, the left vertebral artery was normal, but the state of the right vertebral artery was not known. Her hypertension was difficult to control, and she developed a lupus-like syndrome with hydralazine and unacceptable hirsutism with minoxidil. At age 8 years, repeat renal angiography showed intrarenal arterial abnormalities and renal vein renin ratios were difficult to interpret. Over the next two years her blood pressure remained difficult to control and was raised despite increasing amounts of antihypertensive treatment. She was admitted to hospital at age 10 years with a sudden onset of dysphasia and dysarthria; her cranial bruit which had been getting quieter was no longer audible. Infarction of the left frontal lobe and posterior half of the left hemisphere was detected on computed tomogram. Carotid angiography showed extensive progressive stenoses of both internal carotid arteries with secondary transdural anastomoses. Intracranial-extracranial anastomosis of the left temporal artery to the left internal carotid artery was carried out, distal to the stenotic areas. She remained hypertensive after surgery, requiring a continuous infusion of labetalol. On the third postoperative day, she became more hypertensive and developed increased right sided tone and right divergent gaze. A computed tomogram showed an extensive haematoma in the left parietal region with considerable left to right shift: this was drained and she then made a good recovery. Six months later she was readmitted to hospital having suddenly developed loss of vision. Her blood pressure was 150/95 mmHg, and clinical examination suggested occlusion of the right posterior cerebral artery; this was confirmed by computed tomogram. In view of the haemorrhage into a recent infarction after the first operation, a second bypass was delayed for three months, it was then undertaken uneventfully. Two years later she still has considerable difficulties because of her previous cerebrovascular accidents but she has had no new problems related to vascular insufficiency. Linear growth and pubertal development have been normal. The vascular anastomoses are both pulsatile,

and her blood pressure is controlled at an age appropriate level (120/80 mmHg) on a combination of captopril, propranolol, and frusemide.

Discussion

Some one to three per cent of all children have raised blood pressure. In most the increase is mild, and these children probably fall into the category of essential hypertension.² Ten per cent of them, however, have severe hypertension which is usually secondary in nature, occurring most often in association with the scarred kidneys of vesico-ureteric reflux.³ Between 4.5 and 11.5% of hypertensive children have renal arterial disease, usually some form of renal artery stenosis caused by fibromuscular dysplasia.⁴ The identification of the site and nature of arterial disease may allow treatment either by surgical bypass using autologous saphenous vein grafts or Dacron implants, or more recently, by intraluminal balloon angioplasty.⁵⁻⁷ In some cases, these procedures may result in complete cure and hence obviate the need for lifelong antihypertensive treatment.

Renal arterial disease may be found in association with the neuroectodermal syndromes (for example neurofibromatosis,⁸ Klippel-Trenaunay-Weber,¹ the Feuerstein-Mimms syndromes¹), although the exact incidence of this is as yet unknown. In these patients, particularly, there may be evidence of widespread arterial disease, and neurological symptoms could erroneously be attributed solely to their hypertension.

In the two patients described, there was evidence of neurological impairment; indeed in case 2 this was dramatic and progressive. Angiography in both showed severe, progressive, occlusive intracerebral arterial disease. In case 2, the carotid bruit had resolved and pulsation could no longer be detected in either carotid artery.

The technique of extracranial-intracranial arterial bypass, first described in 1968 by Donaghy and Yasargil,⁹ has been used principally in adults with occlusive vascular disease, and requires anastomosis of the superficial temporal artery to a cortical branch of the middle cerebral artery. As these arteries are about 1 to 1.5 mm in diameter, microsurgery is necessary. The procedure is associated with a low morbidity and mortality in adults, and has produced dramatic improvements.¹⁰

The timing of intervention is critical. The patient in case 2 had suffered a major cerebrovascular accident shortly before her first bypass. At operation, vessels were friable and difficult to suture and afterwards she bled into her previous cerebral infarct, the haematoma requiring surgical evacua-

tion. This complication was probably accentuated by unstable blood pressure after surgery. In the first patient, despite the hemiparesis at age 12 months, no major intracerebral damage had been suffered and there were fewer technical problems. Control of blood pressure after surgery should maintain perfusion through the bypass without provoking cerebral haemorrhage. Maintaining the blood pressure as close to the value before surgery, ideally at one appropriate for the child's age, is probably safest. We found that early reintroduction of oral anti-hypertensive drugs (in the same doses as before operation) was the easiest way of achieving this. Alternatively, intravenous drugs such as labetalol or sodium nitroprusside may be used, but must be introduced as soon as the blood pressure rises.

Both patients had their operations two and a half years ago. The grafts are pulsatile and neither has suffered any further neurological complications. The patient in case 2 remains handicapped by her earlier episodes of vascular insufficiency and attends a special school; possibly earlier intervention might have prevented some of her problems. The first patient is at a normal school but requires some remedial help.

Management of blood pressure has been much easier since the revascularisation procedures in both patients. Before surgery control was often poor, despite multiple drug regimens. The use of captopril before surgery improved control, but in case 1 unmasked his precarious cerebral perfusion. In the kidney, hypoperfusion is associated with a rise in plasma renin activity and hence a rise in blood pressure, retention of sodium increasing circulating blood volume in an attempt to restore normal perfusion. Although renin and renin-like proteins have been isolated from the brain,¹¹ their potential role in the regulation of blood pressure is as yet unclear, it would seem important, however, that blood flow to the brain be maintained at levels adequate to ensure cortical perfusion. It seemed in

our patients that when cerebral blood flow was further impaired blood pressure became even more difficult to control. Since revascularisation both children have been treated with captopril, propranolol, and frusemide. Their blood pressures have been normal, and little manipulation of treatment has been needed.

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