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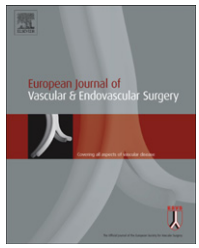

Surgical Treatment and Long-term Outcome of Renovascular Hypertension in Children and Adolescents

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Abstract *Objectives:* This article describes the long-term outcome of surgical treatment in children with renovascular hypertension (RVH) over a 40-year period.
Design: Retrospective study.
Materials and methods: Twenty-five consecutive patients, aged 5–21 years, underwent renal artery (RA) repair from 1967 to 1995. The disease consisted of fibromuscular dysplasia in 17 patients, Takayasu's arteritis in 7 and neurofibromatosis type 1 in one patient.
Results: Twenty-nine RAs were repaired. Primary procedures included aortorenal bypass (ARB) with prosthesis in 10 RAs, autologous vein in five or internal iliac artery in four as conduits, direct reimplantation (DR) in four and nephrectomy in two RAs. Immediate graft failure occurred in three patients despite no peri-operative deaths. After a mean follow-up of 24.4 years, seven patients required secondary nephrectomy. Autologous ARB or DR showed better RA patency and fewer chances for secondary nephrectomy than prosthetic ARB. Hypertension was cured or improved in 21 patients. The overall cumulative survival rate at 20 years was 84%. All five deaths, observed a mean of 12.6 years after the initial operation, were attributed to cardiovascular events.
Conclusions: Surgical treatment, especially autologous ARB or DR, seems to provide durable results for paediatric RVH. Long-term observation and control of hypertension is mandatory.

The overall incidence of hypertension is estimated to be 1–5% of all children in Western countries.[1] Unlike hypertension in adults, a large proportion of paediatric hypertension is potentially correctable. In general, the younger the child and the more severe the hypertension, the more likely a secondary or correctable cause exists. The

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most common cause of surgically correctable hypertension in the paediatric population is renovascular disease.[2] Renal ischaemia can be caused by renal artery stenosis (RAS) or narrowing and coarctation of the aorta or both. A heterogeneous group of arterial diseases is responsible for these lesions.[3] The most frequent cause is fibromuscular dysplasia (FMD). Takayasu's arteritis (TA) is another important cause of paediatric renovascular hypertension (RVH), especially in Asian countries.[4]

In spite of a heterogeneous group of renal artery (RA) lesions, there is a common mechanism whereby renal ischaemia produces persistent elevation of blood pressure. Reduction in RA blood pressure caused by severe RAS leads to hypotension of the kidney and activates the renin–angiotensin system. Excessive release of renin and production of angiotensin II are responsible for systemic hypertension.[5]

Management of RVH consists of proper control of severe hypertension and preservation of the renal parenchyma. RA repairs, whether surgical or endovascular, are essential when hypertension is resistant to anti-hypertensive (AH) medication. This retrospective study examined the efficacy of RA repair in children and adolescents by reviewing the long-term outcomes of 25 consecutive patients in a single institution.

Patients and methods

We reviewed clinical records of patients, 21 years or younger, who were referred to The Second Department of Surgery at The University of Tokyo Hospital for consideration of surgical treatment of RVH. The underlying causes of RA lesions were established by clinical and angiographical evaluation and confirmed by surgical and pathological findings. Diagnosis of TA was confirmed by characteristic clinical and radiological findings[6] that are compatible with the criteria outlined by the American College of Rheumatology.[7] Neurofibromatosis type 1 (NF-1) was diagnosed by multiple *café-au-lait* spots and the presence of a first-degree relative with NF-1, criteria for which were established by the National Institutes of Health.[8] FMD was defined as an idiopathic, segmental, non-inflammatory and non-atherosclerotic disease of the musculature of small- and medium-sized arterial walls including the renal and carotid arteries.[9] The diagnosis was made by radiological and pathological findings, if available.

Patient blood pressure and mortality data were obtained from clinical records, and supplemented with information obtained via telephone interviews with the patients or their physicians. Data obtained during the initial clinical and the final follow-up visit were used for the analysis. Hypertensive retinopathy was positive if retinal findings were classified into Keith–Wagener Grade I or more severe. Plasma renin activity (PRA) was abnormal if it exceeded 2.0 ng ml^{-1} h^{-1} and ipsilateral elevation of PRA was defined as affected side/unaffected side >1.5 or affected side/infrarenal inferior vena cava >1.5. Erythrocyte sedimentation rate (ESR) was evaluated before steroid therapy. Aortography was performed in all patients and selective RA angiography was added to the tests after 1975. RA lesions were analysed based on angiographical findings and were categorised as follows: ost – lesions confined to the ostium of the main trunk; main – lesions confined to the main trunk but not involving the ostium; and seg – lesions of a segmental artery.

Renal artery repair was considered when (1) the diagnosis of RVH was made and other causes of hypertension were ruled out and (2) AH medication failed to control blood pressure.

The RA repair patency was assessed clinically by angiography, duplex scan or both. The RA repair outcome was evaluated based on blood pressure and the requirement for AH medication. Hypertension was defined as average systolic and/or diastolic blood pressure greater than or equal to the 95th percentile for sex, age and height.[10] Blood pressure response was defined as follows:

- Cured – normalisation of blood pressure to below the expected 95th percentile without requiring AH medication;
- Improved – normalisation of blood pressure while on the same drug therapy or decrease in diastolic pressure by more than 15% compared with the preoperative level; or
- Failed – persistent hypertension despite AH therapy with less than a 15% decrease in diastolic pressure compared with the preoperative level.

Results

Patients' background

Patients consisted of 17 females and eight males, whose age at treatment was 17.2 years (range, 5–21 years). FMD was present in 17, TA in seven and NF-1 in one patient. The distribution of patients' ages and aetiologies of RVH are shown in Table 1 and Fig. 1. In principle, all patients had been treated medically for hypertension before admission to our hospital. However, AH medication was abandoned due to its ineffectiveness for blood pressure control in two patients with FMD, and no information regarding medical therapy before admission was available for two patients with FMD. All FMD and two TA patients showed ESR <20 mm h^{-1}, but the remaining five TA patients showed elevated ESR (mean $\pm$ SEM: 80 ± 15 mm h^{-1}). These patients were administered a steroid preoperatively for 31 (range, 3–108) months, during which time ESR levels declined to 12 ± 4 mm h^{-1} at the time of RA repair. One patient with FMD had undergone nephrectomy and another with FMD had recurrent RAS after percutaneous transluminal renal angioplasty (PTRA).

Severe hypertension was present in all patients preoperatively. The mean preoperative blood pressure was $198 \pm 5/111 \pm 3$ mmHg at the time of diagnosis and $182 \pm 6/104 \pm 4$ mmHg on admission with a mean of 2.0 drugs (range, 0–4). Fourteen patients had symptoms or sequelae of severe hypertension including headache, nausea, palpitations and dyspnoea. One patient had seizures. A vascular bruit in the abdomen was audible in 14 patients and hypertensive retinopathy was found in 12. PRA was