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

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KEYWORDS
Renovascular hypertension; Children and adolescents; Renal artery stenosis; Fibromuscular dysplasia; Takayasu’s arteritis; Aorto renal bypass

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.

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most common cause of surgically correctable hypertension in the paediatric population is renovascular disease. 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. 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.

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.

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 that are compatible with the criteria outlined by the American College of Rheumatology. 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. 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. 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. 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 ± 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 ± 5/111 ± 3 mmHg at the time of diagnosis and 182 ± 6/104 ± 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 bruise in the abdomen was audible in 14 patients and hypertensive retinopathy was found in 12. PRA was
estimated in the peripheral blood of all patients and was elevated in 16. Ipsilateral elevation of renin activity was seen in 10 out of 19 patients whose central vein blood was sampled. An abnormal renogram pattern was seen in 21 out of 24 patients who underwent renography. 

Stimulation of renin secretion by administration of captopril, apresoline or furosemide, or by no salt intake was conducted in 12 patients. All PRA and renography data were obtained under AH therapy and with renin stimulation, if present.

Preoperative angiography showed bilateral RA lesions in six, aneurysmal change associated with RA stenosis in five and an aberrant polar RA in seven patients. Locations of the RA lesions are shown in Table 2. All RA lesions in TA were present within the main RA, whereas 32% of those in FMD were located in the segmental arteries.

No patient was affected by aortic coarctation or hypoplasia. Two FMD patients had other vascular lesions—one with stenosis of the subclavian artery and multiple stenoses of pulmonary arteries and the other with moyamoya disease. Four TA and one NF-1 patient exhibited concomitant stenotic lesions of the aortic branches including the innominate (n = 1), subclavian (n = 2), common carotid (n = 1), celiac (n = 2) and superior mesenteric arteries (n = 3). No patient presented with abdominal angina or cerebral ischaemia.

In all cases, RVH was diagnosed based on the presence of persistent hypertension and RAS on arteriography. Arteriography was the only modality for making the diagnosis in two FMD cases, where plasma renin activity and renogram were normal when renin secretion was stimulated.

### RA repair and follow-up

Overall, 29 RAs were repaired in 25 patients (Table 3). Of six bilateral RA lesions, four patients required single-stage reconstruction of the bilateral RAs. The remaining two required single repair, because one patient had already undergone nephrectomy on the other side and the other had only mild stenosis of the contralateral RA. Aorto renal bypasses (ARBs) were performed with prosthetic grafts for 10 RAs and autologous grafts including the saphenous vein and internal iliac artery (IIA) for nine. Use of the IIA was preferred for younger patients but one patient aged 12 years underwent RA bypass with the saphenous vein because the IIA was used for contralateral RA reconstruction. Direct anastomosis of the RA to the aorta after resection of its stenotic lesion (direct reimplantation; DR) was performed for four RAs. Prosthetic bypasses were performed until 1974, while autologous bypass and DR have been performed since 1975. Vein patch angioplasty was performed in two RAs, and PTRA and open operative dilatation were performed in each. Branch reconstruction was required for six patients with FMD or NF-1 and one with TA whose lower segmental artery required patch angioplasty due to a technical problem. All surgical procedures of the affected kidney were performed in situ without using cold saline perfusion. Primary nephrectomy was performed in two patients for whom revascularisation was impossible. No patient required revascularisation of the concomitant stenosis of other aortic branches. Pathological RA examination was performed in 18 patients (11 with FMD and seven with TA), and all microscopical findings were compatible with the clinical diagnosis.

### Short-term outcome (within 30 days after RA repair)

There was no death related to RA repair. An immediate occlusion of reconstructed RA occurred post-operatively in three patients including two with prosthetic ARB and one with patch angioplasty, all of whom underwent branch reconstruction. Another patient with FMD suffered sustained hypertension due to post-operative partial renal infarction despite a patent prosthetic graft. Short-term RA patency was 80% for prosthetic ARB versus 100% for autologous ARB or DR. No patient experienced deterioration of renal function post-operatively. The mean post-operative blood pressure at discharge was $139 \pm 4/80 \pm 3$ mmHg. Hypertension was cured or improved in 22 patients (88%) including one with FMD who underwent unplanned nephrectomy after immediate graft failure.
Control of post-operative blood pressure failed due to graft occlusion in two patients and renal infarction in the other (Table 4).

Long-term outcome (more than 1 month after RA repair)

The follow-up period for 25 patients with RVH ranged from 5 to 39 years (mean ± SEM, 24.4 ± 2.3 years). Although no patient showed vein graft dilatation, four had evident occlusion of the reconstructed RA during the follow-up period. Two patients with TA showed prosthetic graft occlusion in 2 and 8 months after ARB, while another TA patient revealed occlusion of a vein graft 89 months post-operatively. The remaining patient with FMD whose RA was occluded at an unknown time point had severe hypertension and renal failure 12 years after surgery.

Two patients with FMD and four with TA underwent total or partial nephrectomy in the follow-up period. Indications for nephrectomy were recurrent severe hypertension after failure of RA reconstruction in four patients (2 and 8 months after prosthetic ARB in each patient with TA, 8 months after patch angioplasty in a patient with FMD and 89 months after autologous ARB in a patient with TA) and persistent hypertension due to partial renal infarction in a patient with FMD (7 months after prosthetic ARB). The remaining patient in whom the prosthetic graft had immediately occluded developed an anastomotic aneurysm between the aorta and the graft, and successfully underwent aneurysmectomy as well as total nephrectomy 22 years after the initial RA repair. Therefore, 5 out of 10 patients required nephrectomy in the follow-up period after prosthetic ARB while only 1 out of 13 patients required the same after autologous ARB or DR.

Hypertension was cured in 15 (60%), improved in six (24%) and unchanged in four (16%) patients (Table 5). The reasons for poor blood pressure control were failure of the initial RA repair without further surgical treatment for two patients with FMD and one with TA; the reason was unknown for the remaining patient with TA who underwent primary nephrectomy. Thus, blood pressure control was achieved in 15 patients with FMD, five with TA and one with NF-1. These results show no difference in hypertension outcome either between patients with FMD and TA (Table 5), or between patients after prosthetic ARB and those after autologous ARB or DR.

Five deaths occurred at the mean age of 31.8 ± 5.4 years during a mean follow-up period of 12.6 ± 2.8 years (Table 5). All deaths were attributed to cardiovascular events including heart failure in three, myocardial infarction in one, and subarachnoid haemorrhage in one patient. One patient with TA and two with FMD suffered recurrent hypertension after RA reconstruction failure and died of heart failure 5, 9 and 17 years after surgery at age 25, 28 and 36, respectively. The remaining one patient with FMD and one with TA underwent primary nephrectomy at age 20 and 18 years respectively, and died of myocardial infarction and subarachnoid haemorrhage 12 and 20 years, respectively, after the initial surgery. This patient with TA had recurrent hypertension and the other with FMD had well-controlled hypertension. Neither patient had showed RA lesions on the contralateral side at the time of surgery. The cumulative survival rate for all patients was 95%, 90% and 84% at 5, 10 and 20 years, respectively. There was no difference in survival rates between ARB with prosthesis and ARB with autograft/DR (data not shown). In addition, presence of FMD and TA as causes of RAS did not influence the survival rates in our series (Fig. 2).

### Table 2

Vascular lesions.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Number of cases</th>
<th>Number of affected RA</th>
<th>Location of RAS</th>
<th>RAA</th>
<th>Vascular lesions other than RA</th>
</tr>
</thead>
<tbody>
<tr>
<td>FMD</td>
<td>17</td>
<td>19</td>
<td>4</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>TA</td>
<td>7</td>
<td>11</td>
<td>8</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>NF-1</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>


a One with subclavian artery (SCA) stenosis + pulmonary artery multiple stenosis and the other with moyamoya disease.

b One with occlusion of innominate artery + common carotid artery + SCA, one with stenosis of SCA + superior mesenteric artery (SMA), one with stenosis of celiac artery (CA) and occlusion of SMA, one with occlusion of SMA.

c Associated with occlusion of CA.

### Table 3

Initial treatment for RAS.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Number of treated RA</th>
<th>FMD</th>
<th>TA</th>
<th>NF-1</th>
</tr>
</thead>
<tbody>
<tr>
<td>FMD</td>
<td>18</td>
<td>10</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>TA</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>NF-1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Arterial bypass

- **Prosthesis**
  - FMD: 5 cases before Jan 1975, 0 cases after Jan 1975
  - TA: 4 cases before Jan 1975, 1 case after Jan 1975
  - NF-1: 1 case before Jan 1975, 1 case after Jan 1975

- **Vein**
  - FMD: 0 cases before Jan 1975, 0 cases after Jan 1975
  - TA: 0 cases before Jan 1975, 0 cases after Jan 1975
  - NF-1: 1 case before Jan 1975, 1 case after Jan 1975

- **IIA**
  - FMD: 1 case before Jan 1975, 2 cases after Jan 1975
  - TA: 1 case before Jan 1975, 1 case after Jan 1975
  - NF-1: 0 cases before Jan 1975, 1 case after Jan 1975

- **Reimplantation**
  - FMD: 3 cases before Jan 1975, 1 case after Jan 1975
  - TA: 1 case before Jan 1975, 0 cases after Jan 1975
  - NF-1: 1 case before Jan 1975, 0 cases after Jan 1975

- **Patch angioplasty**
  - FMD: 2 cases before Jan 1975, 0 cases after Jan 1975
  - TA: 0 cases before Jan 1975, 0 cases after Jan 1975
  - NF-1: 2 cases before Jan 1975, 0 cases after Jan 1975

- **Angioplasty**
  - FMD: 0 cases before Jan 1975, 0 cases after Jan 1975
  - TA: 0 cases before Jan 1975, 0 cases after Jan 1975
  - NF-1: 0 cases before Jan 1975, 0 cases after Jan 1975

- **Primary nephrectomy**
  - FMD: 1 case before Jan 1975, 1 case after Jan 1975
  - TA: 1 case before Jan 1975, 0 cases after Jan 1975
  - NF-1: 0 cases before Jan 1975, 1 case after Jan 1975

FMD: fibromuscular dysplasia, TA: Takayasu’s arteritis, NF-1: neurofibromatosis type 1, RA: renal artery, IIA: internal iliac artery.

a Cases before Jan 1975.
b Cases after Jan 1975.
Discussion

This article reviewed the surgical treatment for RVH in children and adolescents with heterogeneous types of RA lesions. Similar to other papers describing paediatric RVH, our study revealed that FMD was the most common cause of RAS. A pathological classification of renal artery FMD was based on a dominant arterial wall layer that shows dysplasia or hyperplasia, and lesions are classified as three main types: intimal, medial and perimedial. Medial and perimedial FMD account for nearly 95% of all FMD stenosis cases. TA, characterised by non-specific inflammation of the aorta and its branches, is a major cause of RVH in the young population, especially in Asian countries. Thus, unlike Western countries, a high incidence of TA was seen in our series. Most of our patients with TA had an ostial RA lesion, in contrast to FMD, in which not only the main artery but its segments are often affected.

NF-1 is the most common cause of genetically determined RA disorders. Vascular lesions associated with NF-1 usually consist of stenosis of the aortic branches, especially the RA, SMA and CA, and coarctation of the abdominal aorta.

The diagnostic pathway of RVH in children was shown in a recent review. Children showing signs of RVH such as severe hypertension, abdominal bruises, end-organ damages and raised peripheral PRA, as well as those who are at a high risk of vascular disease, should undergo investigation including angiography and central vein renin sampling. The RVH treatment pathway shows that children with blood pressure that is not well controlled with more than two drugs should receive RA revascularisation, which aims to control blood pressure and preserve renal function. In our and other paediatric cases, RA revascularisation had favourable clinical outcomes. However, in a recent randomised trial, RA revascularisation for atherosclerotic RAS had no or little benefit for renal function, blood pressure control, morbidity and mortality. This discrepancy may indicate that RAS in young patients shows rather isolated lesions and plays a major role in blood pressure control and renal function, compared with RAS in atherosclerosis.

**Table 4** Short-term outcome.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>FMD</th>
<th>TA</th>
<th>NF-1</th>
<th>Procedures</th>
<th>Prosthetic ARB</th>
<th>Autologous ARB/DR</th>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>17</td>
<td>7</td>
<td>1</td>
<td>9 (10)</td>
<td>2</td>
<td>0</td>
<td>6 (6)</td>
</tr>
<tr>
<td>Graft/patch failure</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Secondary nephrectomy</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Short-term outcome</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cured</td>
<td>10</td>
<td>5</td>
<td>1</td>
<td>5</td>
<td>9</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Improved</td>
<td>5</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>2a</td>
<td>3a</td>
<td>3a</td>
</tr>
<tr>
<td>Failed</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

FMD: fibromuscular dysplasia, TA: Takayasu’s arteritis, NF-1: neurofibromatosis type 1, ARB: aortorenal bypass, DR: direct reimplantation.

The number of repaired RA is shown in parentheses.

**Table 5** Long-term outcome.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>FMD</th>
<th>TA</th>
<th>NF-1</th>
<th>Procedures</th>
<th>Prosthetic ARB</th>
<th>Autologous ARB/DR</th>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>17</td>
<td>7</td>
<td>1</td>
<td>9 (10)</td>
<td>11 (13)a</td>
<td>6 (6)a</td>
<td>6 (6)a</td>
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<tr>
<td>Graft/patch failure</td>
<td>1</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Secondary nephrectomy</td>
<td>2</td>
<td>4</td>
<td>0</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Long-term outcome</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Cured</td>
<td>12</td>
<td>3</td>
<td>0</td>
<td>4</td>
<td>9</td>
<td>2</td>
<td>2</td>
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<tr>
<td>Improved</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Failed</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1a</td>
<td>3a</td>
<td>3a</td>
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<tr>
<td>Death</td>
<td>3</td>
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<td>0</td>
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<td>1a</td>
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<tr>
<td>Congestive heart failure</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>2a</td>
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<td>2a</td>
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<tr>
<td>Myocardial infarction</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Subarachnoid haemorrhage</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

FMD: fibromuscular dysplasia, TA: Takayasu’s arteritis, NF-1: neurofibromatosis type 1, ARB: aortorenal bypass, DR: direct reimplantation.

The number of repaired RA is shown in parentheses.

a Both numbers include a patient with FMD who underwent autologous ARB for the right and angioplasty for the left RA, and died of heart failure.
Therefore, aggressive treatment of RAS should be advocated for paediatric RVH.

We performed prosthetic RA revascularisation prior to 1975, but changed to RA bypass with autologous RA bypasses or DR thereafter. This shift was justified because autologous ARB or DR improved RA patency, at least within 1 year after surgery, and lessened the chance for secondary nephrectomy. Use of IIA grafts as a conduit for autologous bypasses is preferred, because its patency is good and graft dilatation is rare. For ostial lesions, DR is also a durable alternative with little chance of anastomotic complications. In contrast to autologous grafting, use of prosthesis might lead to failure of the anastomosis or graft occlusion since the prosthesis does not change in size along with normal vessel growth and the amount of RA blood flow may be too small for long-term patency of small-calibre prosthetic grafts. In spite of better patency, autologous ARB or DR showed no difference in long-term blood pressure control compared with prosthetic ARB. This may be explained by the fact that secondary nephrectomy improved hypertension control in five patients whose prosthetic grafting failed. Thus, nephrectomy can be an acceptable treatment option after RA revascularisation failure. Immediate failure of RA repair was seen only in patients undergoing RA branch reconstruction with prosthesis or vein patch angioplasty, suggesting that not only the kind of conduit but also the technique may be critical for graft patency.

Recently, there have been a growing number of PTRA experiences for the treatment of paediatric RAS. Etiology and the type of lesion are particularly important when PTRA is considered. As lesions in FMD sometimes affect RA branches, the surgical procedures are often complicated, so PTRA may be a better solution. Primary patency after PTRA is more than 80% at 10 years, and PTRA can be repeated if re-stenosis occurs. The efficacy of PTRA in TA and NF-1 patients is more disappointing and its performance is still controversial. The main reason for only one patient undergoing PTRA in our series was the high prevalence of surgical procedures and the low availability of PTRA during or prior to the 1990s. Recent advances in endovascular techniques enable many cases with RAS to be treated with PTRA, but it should be noted that surgical treatment is still indicated, especially for lesions such as extensive RA involvement, aneurysms and atrophic kidney.

Table 6 lists recent reports on hypertension outcomes after surgical treatment for RVH in 25 or more children and adolescents. All of these reports demonstrated that FMD was a prominent etiology of RA lesions, and more than 85% of the surgical procedures were RA reconstructions that included bypass or DR. Long-term hypertension outcomes were similar among these reports, and approximately 90% or more of patients benefited from surgical treatment. Only our study has a mean follow-up period of 24.4 years, while the others showed a mean of less than 10 years. It is preferable for paediatric patients to be followed up for more than a decade when the long-term outcome of RA repair is evaluated.

We have tried to examine the difference in blood pressure response between patients with FMD and TA despite the small study population. Very few articles have described this difference. Failure rates of RA repair depend not on the etiology of RA lesions but on the procedure of RA repair, as shown by inferior patency of prosthetic grafting, at least those within a year after surgery. Furthermore, RA repair including secondary nephrectomy benefited 15 out of 17 patients with FMD and five out of seven with TA during the follow-up period, showing no significant difference in blood pressure responses between patients with FMD and TA in the long-term period.

Causes of death after surgical treatment of RVH were not discussed in either report shown in Table 6. Five patients undergoing PTRA in our series was the high prevalence of surgical procedures and the low availability of PTRA during or prior to the 1990s. Recent advances in endovascular techniques enable many cases with RAS to be treated with PTRA, but it should be noted that surgical treatment is still indicated, especially for lesions such as extensive RA involvement, aneurysms and atrophic kidney.

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Table 6  Reports on hypertension outcomes after surgical treatment of renovascular hypertension in children and adolescents.

<table>
<thead>
<tr>
<th>Location</th>
<th>N</th>
<th>Years</th>
<th>Hypertension outcome</th>
<th>F/U year range (Mean)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>% Cured</td>
<td>% Improved</td>
</tr>
<tr>
<td>Ann Arbor, Mich</td>
<td>97</td>
<td>1963–2006</td>
<td>70</td>
<td>27</td>
</tr>
<tr>
<td>Clincy, France</td>
<td>78</td>
<td>1975–98</td>
<td>87</td>
<td>10</td>
</tr>
<tr>
<td>Cleveland, Ohio</td>
<td>56</td>
<td>1955–88</td>
<td>66</td>
<td>23</td>
</tr>
<tr>
<td>Nashville, Tenn</td>
<td>53</td>
<td>NS</td>
<td>70</td>
<td>26</td>
</tr>
<tr>
<td>Philadelphia, Penn</td>
<td>25</td>
<td>1987–2004</td>
<td>36</td>
<td>56</td>
</tr>
<tr>
<td>Tokyo, Japan</td>
<td>25</td>
<td>1967–95</td>
<td>60</td>
<td>28</td>
</tr>
</tbody>
</table>

N: number of patients, F/U: follow-up, NS: not stated.
deaths occurred during a mean follow-up period of 12.6 years in our series, and all deaths were cardiovascular-related. Four out of five patients who died suffered from hypertension, and this condition might have been related to the fatal events. As shown in Fig. 2, there is no difference in survival rates between patients with FMD and TA. It is interesting to note that a patient with TA who underwent primary nephrectomy developed severe hypertension, probably due to stenosis of the contralateral RA, which was patent at the time of the initial surgery. This indicates that long-term close follow-up is essential in terms of proper hypertension control and scrutiny of RA patency of both the affected and the contralateral side.

In conclusion, we have examined RA repair in 25 paediatric and adolescent cases with RVH whose aetiologies included FMD in 68% and TA in 28%. We have shifted our strategy of RA revascularisation from prosthetic ARB to autologous ARB or DR. This has led to the improvement of RA patency and a decrease in the number of cases requiring secondary nephrectomy. Secondary nephrectomy might be an alternative for patients who lack hypertension control due to graft failure. Because all deaths were attributed to cardiovascular events, long-term follow-up is mandatory especially to ensure hypertension control.

**Conflict of Interest/Funding**

None

**References**