Surgical Treatment of Renovascular Hypertension in Children

M. Lacombe*

Consultation de Chirurgie, Hôpital Beaujon, 92119 Clichy Cedex, France

Submitted 4 January 2011; accepted 14 February 2011
Available online 21 March 2011

Abstract

Objectives: The aim of this retrospective study was to report the author's experience of the surgical treatment of renovascular hypertension in children and to define the role of surgery in its treatment.

Material and methods: This series includes 85 patients (50 girls, 35 boys), 28 months to 18 years of age (mean: 10.3) operated on from 1970 to 2005. All patients had arterial hypertension and underwent the investigations usually performed in hypertensive patients. Renal artery lesions were bilateral in 26 cases. Due to bilateral procedures and to secondary or late reoperations, the number of surgical procedures was 114 (15 nephrectomies and 99 arterial repairs).

Results: Fibrodysplasia of the renal artery was the prevailing pathologic factor (71%). Associated vascular lesions were observed in 61% of the patients. There was no postoperative death in this series. Seven postoperative thromboses occurred (7% of the repairs). The complete cure of arterial hypertension was obtained in 82% of the patients. In young children, growth of the repairs was normal when age increased.

Conclusion: Surgery still holds a prominent place in the treatment of renovascular hypertension in children. Its prognosis is favourable since atheroma or organ lesions are usually lacking.

© 2011 European Society for Vascular Surgery. Published by Elsevier Ltd. All rights reserved.

Renal artery lesions are the major causes of arterial hypertension in children. According to previous publications, their frequency ranges from 6% to 20% in hypertensive children.1,2 They differ from the lesions observed in adult patients. In the latter, atheroma is the prevailing cause of renal artery disease, accounting for 70–80% of all cases, whereas fibrodysplasias are predominant in the paediatric population. Moreover, most patients with atheromatous renal artery stenoses have associated atherosclerosis of other arterial territories, whereas in children the arterial system remains unaltered. The diagnosis of renal artery lesions is especially important, since their surgical repair or their treatment by percutaneous transluminal renal angioplasty (PTRA) can cure the arterial hypertension. The purpose of this article is to define the role of surgery in the treatment of renal artery lesions.
Patients and Methods

Patient population

This is a retrospective study of all children operated by the author for lesions of the renal artery(ies). They were operated between 1970 and 2005, date at which the study was ended in order to have a follow-up of at least 5 years for all patients. These children were referred to the author by different centres of paediatric nephrology in France and Europe. Among the 87 patients of this series, two died suddenly before any treatment and 85 were treated, of whom 50 were girls and 35 boys. The upper age limit of these patients was 18, which is the age of legal majority in France. Their ages ranged from 28 months to 18 years (mean: 10.3 \pm 5.8).

Preoperative check-up

The presence of a lesion of a renal artery was recognised by the usual investigations. The duplex ultrasound was the basic investigation for the detection of these lesions. The main criteria to confirm stenoses were the changes of spectral waveform (spectral broadening, systolic peak velocity > 180 cm s\(^{-1}\), turbulent flow) and, with colour imaging, the direct visualisation of the renal arteries. Arterial opacification was performed by arteriography using the Seldinger’s technique in the first years of the author’s experience and by intra-arterial digital angiography since the 1980s. An increasing number of patients were investigated by angio-computed tomography (CT) scan and angio-magnetic resonance imaging (MRI) during the last decade.

The consequences of the elevated blood pressure were appreciated in the same way as in adult patients. The renal function was evaluated by the measures of blood creatinine and creatinine clearance. Sometimes, the function of each kidney was assessed separately by renal scintigraphy. The cardiac status was evaluated by electro- and echocardiography. The consequences of arterial hypertension on the nervous system were evaluated by examination of the fundus.

Investigations suggesting the renovascular cause of arterial hypertension were rarely used in children: selective renal vein sampling is invasive and the tests using angiotensin inhibitors or angiotensin-converting enzyme inhibitors require a strict methodology in experienced centres.

Surgical treatment

Due to bilateral procedures and to secondary or late reoperations, the number of surgical procedures was 114 (15 nephrectomies and 99 arterial repairs). The techniques of renal artery repair are shown in Table 1. Bilateral lesions were present in 28 patients (33%) who underwent a repair on both sides and 14 repairs (14%) were performed on solitary kidneys. Altogether, there were 52 lesions on the right side and 59 on the left side.

Arterial substitutes for the renal artery reconstruction were used in 53 repairs (53% of them): 48 internal iliac artery autografts and five autogenous saphenous vein grafts. Synthetic prostheses were used only for aortic bypasses and not for renal artery repair.

Single sutures were used whenever non-absorbable material was employed. When absorbable monofilament sutures became available, continuous sutures became of routine use.

In 19 patients (22%), other vascular procedures were by the renal artery repair: superior mesenteric artery repair in seven patients, aortic prosthetic bypass in six, resection of aneurysms of splanchnic arteries in three, removal of pheochromocytomas in two (with three tumours) and section of an arcuate ligament in one.

Postoperative follow-up

Before leaving the hospital, the majority of operated patients had a control digital angiography. Due to its invasive character, this investigation was not performed in patients under 5 years of age who had only duplex colour ultrasound.

In all patients, angiographic investigations were repeated 5 years after surgery to study the evolution of the arterial repair with body growth. Follow-up time ranged from 1 to 18 years (mean: 9.4 years).

Results

Renal artery lesions

All these patients had a stenosis of a renal artery, usually located on the main trunk of the artery. Stenoses of terminal branches were generally associated with a stenosis of the main trunk of the artery. Aneurysms of the renal

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Techniques of renal artery repair.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Segmental resection and anastomosis (± graft interposition)</td>
<td>7</td>
</tr>
<tr>
<td>Aortic reimplantation of the renal artery:</td>
<td></td>
</tr>
<tr>
<td>direct reimplantation</td>
<td>17</td>
</tr>
<tr>
<td>with graft interposition</td>
<td>23</td>
</tr>
<tr>
<td>Anastomosis with splanchnic arteries:</td>
<td></td>
</tr>
<tr>
<td>splenorenal anastomosis</td>
<td>22</td>
</tr>
<tr>
<td>hepatorenal or gastroduodenorenal anastomosis</td>
<td>5</td>
</tr>
<tr>
<td>Extracorporeal surgery with kidney reimplantation or autotransplantation</td>
<td>25</td>
</tr>
<tr>
<td>Total(^a)</td>
<td>99</td>
</tr>
</tbody>
</table>

\(^a\) Number of repairs.
artery were associated with the stenosis (Fig. 1) in 10 patients (11 aneurysms). All these lesions were treated during the same surgical procedure.

The nature of the arterial lesions is shown in Table 2. Due to the difficulty in removing the abnormal artery in several cases, pathologic examination could not be performed in 25 patients. In these cases, the nature of the arterial lesion could only be hypothesised. Fibrodysplasia of the renal artery was the most frequent pathologic finding (71%). Segmental stenoses were observed in 87 cases and extensive ones in 27 cases.

In seven cases (8%), the aspect was that of a congenital hypoplasia or atresia (Fig. 2). These lesions were located on the origin of the renal artery. They differed from segmental dysplastic stenoses by their external aspect resembling a narrow cord, their association with a sometimes huge post-stenotic dilation and their histologic aspect including thinning of the layers of the artery with sparse fibrous lesions and fragmentation of the elastic fibres.

Complete obstruction of the renal artery did not necessarily entail the destruction of the kidney. Frequently, a rich collateral circulation maintained the viability of the organ and it was possible to find distally a renal trunk which could be revascularised.

Takayasu’s disease (Fig. 3) was observed in eight patients (9%). The origin of the patients, coming from East Asia or from countries around the Mediterranean Sea, the presence of an inflammatory syndrome and the association of lesions in other vascular territories such as supra-aortic trunks or pulmonary arteries were highly suggestive of this diagnosis.

Recklinghausen’s neurofibromatosis was present in four patients (5%): specific lesions interesting the renal artery were observed in only two of them and medial fibrodysplasia in the remaining two others.

Other causes (dystrophy of elastic tissue and post-traumatic fibrosis) were far less frequent.

Kidney lesions

Histologic lesions of the kidney were studied in the 1970s on a small number of renal biopsies to obtain some prognostic argument about the evolution of hypertension after surgical repair of the lesions. We rapidly gave up this practice, since this investigation frequently showed only signs of parenchymal ischaemia without other lesions or very discrete ones. This absence of severe lesions was considered compatible with the concept of ‘protection’ of the kidney by

| Table 2 Nature of operated lesions of the renal artery. |
|-----------------|---------------|---------------|
| Nature of the lesion | Certaina (pathology) | Probablea (no pathology) |
| Fibrodysplasia | | |
| segmental | 34 | 11 |
| extensive | 14 | 2 |
| Chronic obstruction (+thrombosis) | 5 | – |
| Recklinghausen’s neurofibromatosis | 2 | – |
| Congenital hypoplasia, atresia | – | 7 |
| Inflammatory arteritis (Takayasu’s disease) | 3 | 5 |
| Dystrophy of elastic tissue | 1 | – |
| Post-traumatic fibrosis | 1 | – |
| Totala | 60 | 25 |

– None.

a Number of patients.
the stenosis. When the kidney appeared severely altered on preoperative investigations, primary nephrectomy was carried out. In such cases, the microscopic examination always revealed severe lesions: glomerular sclerosis, interstitial fibrosis with tubular atrophy, arteriolar fibrosis, subendothelial fibrinoid deposits and large segmental infarcts.

Associated lesions

These were especially frequent in children since they were observed in 52 patients of the series (61%), for a total of 59 lesions, seven patients having multiple associated lesions. These lesions are shown in Table 3.

Circumstances of discovery

In 85% of the patients, the arterial hypertension was clinically silent and was initially overlooked until a medical examination performed for any other medical reason (infectious episode, vaccination, etc.) discovered fortuitously the elevated blood pressure. Severe or decompenated forms were observed in 15% of the patients. As previously mentioned, two children died before any treatment: one from cerebral haemorrhage and the other from acute cardiac failure. In four children, the initial clinical picture was that of a hypertensive encephalopathy with signs of intracranial hypertension and even coma. These symptoms regressed after urgent anti-hypertensive treatment. Five patients had a major hypertensive cardiopathy with frequent pulmonary oedema and with or without cardiac rhythm disturbances, which required correction before surgery. Two children had an emergency surgery because of an acute thrombosis of the renal artery due to the anti-hypertensive treatment.

Mortality, morbidity

There was no postoperative mortality in this series.

Six patients (7%) had a general complication: one transient oliguria after a complex arterial repair on a solitary kidney, three postoperative haemorrhages (two reoperations and one spontaneously resolutive haematoma), one venous iliac thrombosis treated by anticoagulants and one sub-acute rupture of a cerebral aneurysm cured after emergency neurosurgical operation.

Arterial complications were observed in 10 patients (12%): seven occlusions of the renal artery repair occurred, four of which required a secondary nephrectomy; two occlusions of secondary renal branches were responsible for segmental renal atrophy; and one occlusion of a repair of the mesenteric superior artery remained clinically silent due to the compensation by a large Riolan’s arch.

Anatomical results

Five iterative stenoses of the renal artery (5% of all repairs) occurred: three were successfully treated by ex situ surgery, one was treated by transluminal angioplasty and the last received no treatment because the stenosis was moderate and did not entail arterial hypertension. Two aneurysms occurring on venous autografts were resected and replaced by an arterial autograft. In total, the rate of good anatomical results was 89%.

Results on blood pressure status

After the operation, the blood pressure became normal in a few days. However, sometimes, it lowered only progressively in the following weeks up to 3 months. The reasons for this delay are not explained. With a follow-up from 1 to 18 years, 68 patients (82%) have a normal blood pressure (systolic pressure <120 mmHg, diastolic pressure <70 mmHg) and do not follow any diet or receive any anti-hypertensive treatment. Ten children (12%) remain hypertensive but are greatly improved with a normal blood pressure under a moderate medical therapy. In five children (6%) the surgical treatment failed requiring the continuation of a heavy medical treatment. In three of the latter an anatomical factor seems...
responsible (remaining stenosis of the abdominal aorta, stenosis of the coeliac trunk above a splenorenal anastomosis); in the two others, parenchymatous lesions of the kidney have been identified on renal biopsies.

**Long-term evolution**

Late angiographic investigations constantly showed a normal growth of the arterial autografts, of all vascular anastomoses and of the size of the revascularised kidney with ages of the children.

In two children, a stenosis of the opposite renal artery occurred at 1 and 3 years after the initial operation. Both underwent successful surgical repair.

In two patients, a congenital stenosis of the abdominal aorta worsened 3 and 12 years after the initial operation with recurrence of arterial hypertension. The worsening of the aortic lesion was considered responsible for the elevated blood pressure since both patients had undergone a bilateral renal autotransplantation in the iliac fossa and since angiography had shown normal patency of the renal repairs. These two patients underwent a prosthetic aortic bypass which cured the arterial hypertension.

Two patients with Takayasu’s disease developed a lesion in a distal territory: one coronary lesion treated by a coronary bypass 8 years after the renal artery repair and one carotid stenosis which required two successive repairs.

**Discussion**

Renal artery disease is second in frequency to coarctation of the thoracic aorta as the most common form of surgically correctable hypertension in the paediatric population.

---

**Table 3**  Associated lesions to renal artery stenosis in children.

<table>
<thead>
<tr>
<th>Nature of the lesion</th>
<th>Number of lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal aorta</td>
<td></td>
</tr>
<tr>
<td>congenital malformative stenosis</td>
<td>18</td>
</tr>
<tr>
<td>inflammatory stenosis (Takayasu’s disease)</td>
<td>6</td>
</tr>
<tr>
<td>dysplastic aneurysm</td>
<td>1</td>
</tr>
<tr>
<td>Coeliac trunk</td>
<td></td>
</tr>
<tr>
<td>obstruction</td>
<td>5</td>
</tr>
<tr>
<td>compression by a median arcuate ligament</td>
<td>3</td>
</tr>
<tr>
<td>pancreaticoduodenal aneurysm</td>
<td>1</td>
</tr>
<tr>
<td>Mesenteric superior artery</td>
<td></td>
</tr>
<tr>
<td>stenosis or obstruction</td>
<td>12</td>
</tr>
<tr>
<td>aneurysm</td>
<td>2</td>
</tr>
<tr>
<td>Distant arterial territories</td>
<td></td>
</tr>
<tr>
<td>thrombosis of the subclavian artery</td>
<td>1</td>
</tr>
<tr>
<td>aneurysm of the subclavian artery</td>
<td>2</td>
</tr>
<tr>
<td>stenosis of pulmonary arteries</td>
<td>2</td>
</tr>
<tr>
<td>aortic valvular lesion</td>
<td>2</td>
</tr>
<tr>
<td>cerebral aneurysm</td>
<td>1</td>
</tr>
<tr>
<td>Pheochromocytoma</td>
<td>3</td>
</tr>
</tbody>
</table>

---

**Figure 4**  Left: congenital malformative stenosis of the abdominal aorta, stenosis of the left renal artery, atresia of the main trunk of the right renal artery (10 year-old girl). Right: congenital aneurysm of the abdominal aorta, congenital absence of the left kidney, stenosis of the right renal artery (4 year-old girl).
The clinical features of renovascular hypertension are remarkable since the tolerance of the elevated blood pressure may extend over a long period of time but, as was already mentioned by Broyer et al., its decompensation may be extremely fast, sometimes within a few hours. In our series, the two deaths occurring before any treatment and the rate of severe forms attest this rapid evolution. By contrast, cardiomegaly and lesions of the fundus regress very quickly after the operation.

The lesions of the renal artery are mostly congenital. Their association with anomalies of the aorta and its branches (Fig. 4) is in favour of a regional congenital malformation of these arteries. In the 1930s, Maycock had already questioned the role of an anomaly of fusion of the embryonic dorsal aortas to explain the origin of these lesions. An accelerated atrophy or a trouble of growth of one dorsal aorta affecting its branches may be responsible for such association. Similarly, lesions of atresia or hypoplasia of the renal artery, developmental renal artery stenoses according to Stanley, seem to be due to an arrest of growth of the vessel. These problems of growth are sometimes in relation with a viral infection, especially by the rubella virus, during pregnancy, several viruses having a cytotoxic effect which block the aortic growth.

If latent infection, for instance after umbilical catheterisation (performed in two patients of this series), might be the cause of the aortic lesion, remains an open question. The presence of pheochromocytomas and of Recklinghausen’s neurofibromatosis in several patients suggests an affection of the elements of the neural crest and questions about the genetic origin of some renal artery fibrodysplasias.

The diagnosis of these lesions may be suspected on clinical data: the presence of an abdominal systolic bruit, the resistance to medical therapy are highly suggestive. Among the morphologic diagnostic investigations, the echo-Doppler still keeps a prominent place. During the last decades, the considerable progress of vascular imaging techniques has allowed angio-scans and angio-MRI to replace angiography because of its invasive character.

Especially in young children, these new techniques are now routinely used.

The treatment of renal artery lesions includes three complementary methods.

The medical treatment is usually the first applied to hypertensive patients, especially in severe cases. However, it should be remembered that renovascular hypertension is sometimes resistant to anti-hypertensive therapy. Moreover, in tight stenoses, the medical treatment may be responsible for an acute obstruction of the renal artery. In addition, the continuation of this treatment in the long term may be responsible for an irreversible atrophy of the kidney as a result of the permanent reduction of the renal arterial flow due to the stenosis and to the lowering of the arterial pressure. For these reasons, the indications of the medical treatment are limited to moderately hypertensive children in the absence of a tight stenosis. In children under 2 years of age, the medical treatment alone may be useful while waiting for the patient to be old enough for more compatibility with surgical repair.

PTRA has taken a prominent place in the treatment of renal artery stenoses since the 1980s and it is often performed as the initial therapeutic procedure. However, indications of PTTRA in children must be closely discussed. First, angioplasty is not always technically feasible: when the stenosis is very tight or when the arterial lumen is very tortuous as in some extensive fibrodysplasias, catheterisation of the artery may be impossible. Second, angioplasty may be unsuccessful: in severely fibrotic lesions or in cases of developmental stenosis, the lesion will not respond even to high-pressure inflation of the balloon and the use of cutting balloons in such cases may be risky. Third, angioplasty may be responsible for severe complications: arterial dissection, rupture, aneurysm, thrombosis and renal infarct. Shroff et al. observed one death in relation with PTRA. Fourth, all series mention a restenosis rate of up to 20–30%, which requires a prolonged follow-up of the patients. Fifth, the presence of a dysplastic aneurysm associated to the stenosis is a contraindication of angioplasty due to the risk of aneurismatic rupture.

For all these reasons, PTRA has a very limited place in the treatment of renal artery lesions in children. Its indications must be reserved to very young severely hypertensive children as a waiting solution to an age and a size

Figure 5   Left: hypoplasia of the abdominal aorta with bilateral stenosis of the renal arteries in a 3 year-old girl. Right: sequential aorta-to-both renal arteries bypass using a segment of Riolan’s arch. With a follow-up of 18 years, the blood pressure is normal; there is no need of an aortic repair.
compatible with surgery or to restenoses after surgical repair (one successful case in the current series). Stents must be avoided in children because their behaviour is unknown after several decades of time.

In the current series, 16 patients underwent angioplasty in their medical centres before being referred to the surgical unit. The failures, complications, incomplete results and restenoses justified the surgical repair. Four children were declined for PTRA by the interventional radiologist himself.

In case of failure, repeat PTRA is to be avoided because it may worsen the lesions of the renal artery and make surgery much more difficult. It must also be avoided in children with solitary kidneys due to the risk of severe complications of this procedure.

'The surgical treatment' appears, from this large experience and from other recent publications, to have always a prominent place in the care of children with renovascular hypertension. In this series, the number of operations performed was two to three per year until the 1990s. It then increased to six to eight per year.

In the recent years, operated patients had more severe or complex lesions, as compared to patients operated during the 1970s, and required consequently a greater proportion of extracorporeal surgery.

Nephrectomy was necessary in 15 (18%) of the operated children. Severe lesions of the renal parenchyma were the prevailing reason for nephrectomy in 11 of them. Occlusion of the renal artery repair required nephrectomy in four.

Renal artery repair is the ideal treatment. Several techniques may be used (Table 1); however, those avoiding the use of arterial substitutes are preferable. Whenever substitutes are necessary, arterial autografts are the material of choice since they do not undergo late anatomical degenerative changes such as venous autografts or synthetic prostheses. Extracorporeal surgery is reserved to particularly complex lesions.

It is remarkable to observe that, in the long-term follow-up of these patients, the growth of arterial autografts and of vascular anastomoses followed that of the body.

Whenever possible, associated arterial lesions should be repaired during the same procedure. Only complex and multiple lesions may require two separate operations, which was the case for six children of this series (7%). However, associated lesions do not always require immediate repair. Aortic stenoses may be left intact in very young children (Fig. 5) or when they are moderate and/or asymptomatic. When the aorta seems very narrow, its repair is preferably postponed until the age of puberty or of end of growth (Fig. 6). Stenoses of arteries supplying the digestive tract are often asymptomatic since they are compensated by a huge Riolan’s arch and do not require repair; only stenoses of both coeliac trunk and superior mesenteric artery or of the three digestive arteries and aneurysms in these territories are indications for repair. In Takayasu’s disease, because of its evolutivity, arterial repairs should preferably be performed when the inflammation tests are normalised.

'The prognosis' of renovascular hypertension is favourable in children. The short duration of the elevated blood pressure, the absence of atheromatous changes of the arterial system and the usual lack of renal or visceral lesions secondary to hypertension explain the good results of renal artery repair with respect to control of hypertension and preservation of renal function. The long-term results show that the repairs are durable with time and support an aggressive surgical approach towards renovascular hypertension for the paediatric patient.

Conflict of Interest

None.
Funding

None.

References