

Renovascular Diseases

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Summary

This chapter reviews the diagnosis and treatment of surgically correctable hypertension due to renal artery stenosis and aneurysms. The differential diagnosis of secondary hypertension includes adrenal diseases which are discussed in Core Curriculum [Adrenal Neoplasms](#) and [Kidney, Adrenal, Ureter Anatomy](#).

1. Introduction

The most common chronic medical condition in the world is high blood pressure.^{1,2} It is estimated that approximately 33% of Americans have hypertension and the prevalence is expected to increase by another 7.2% by 2030.² Hypertension is associated with substantial cardiovascular morbidity and mortality.³ Secondary hypertension has an identifiable and potentially reversible etiology. There is now significant consensus on the appropriate diagnosis and treatment of hypertension.⁴ It is estimated that 10% to 15% of adults and up to 85% of children have secondary hypertension. Secondary hypertension should be suspected in any patient younger than 30 years without a family history of hypertension or obesity, patients with rapid onset or accelerated high blood pressure and patients with refractory hypertension. (see [Table 1](#))⁵

This chapter will focus on renovascular diseases which include a variety of conditions that affect the renal arterial circulation. Although disorders of the renal vein(s) could correctly be considered renovascular diseases, herein we focus on arterial disorders as primary renal vein disease is far less common. The preponderance of renovascular diseases comprise stenoses of the renal artery resulting from heterogeneous conditions including **atherosclerosis, fibrous dysplasia, vasculitis, neurofibromatosis, congenital bands, intrinsic occlusion from endovascular devices, and extrinsic compression from neoplasia and radiation** ([Table 2](#)). Renal artery stenosis (RAS) has been shown to cause severe hypertension and renal parenchymal loss and revascularization can lead to improvements in both blood pressure control and renal function. Aneurysms of the main or branch renal arteries are also renovascular diseases.

1.1 Keywords

Hypertension, renal artery stenosis, renovascular disease, atherosclerosis, fibrous dysplasia, renal artery aneurysm.

Table 1. Age based approach to the differential diagnosis of secondary causes of hypertension

Children	Age 19 - 29	Age 30 - 64	Older Adult
Congenital anomalies	Thyroid disease	Hyperaldosterone	Chronic kidney disease
Adrenal disease	Fibromuscular dysplasia	Pheochromocytoma	Renovascular disease
Glomerular disease	Chronic kidney disease	Hypercortisol	Obstructive uropathy

Table 2. Etiologies and relative proportion of renal artery diseases

Atherosclerosis	~ 90%
Fibrous dysplasia	~ 10%
Vasculitis	~ 1%
Congenital bands	~ 1%
Intrinsic occlusion (endovascular device malfunction)	~ 1%
Extrinsic compression (neoplasia/radiation)	~ 1%

A retrospective analysis was performed of consecutive pediatric patients with renovascular hypertension at the University of Michigan from 1991 to 2017. One hundred sixty-nine children (76 girls, 93 boys) underwent primary index operations at a median age of 8.3 years. Thirty-one children (18%) had neurofibromatosis type I, 76 (45%) had abdominal aortic coarctations, and 28 (17%) had a single functioning kidney. The overall experience revealed hypertension to be cured in 74 children (44%), improved in 78 (46%) and unchanged in 17 (10%). Children undergoing remedial operations were less likely (33%) to be cured of hypertension.⁶

2. Atherosclerotic Renal Artery Stenosis

Atherosclerotic renal artery stenosis (ARAS) is a recognized cause of severe, secondary hypertension and impaired renal function. Whether ARAS is causal or is an associated finding remains unclear; however, the presence of ARAS is an independent predictor for adverse cardiovascular events and increased cardiovascular mortality.^{7,8} Moreover, the increased risk profile appears to remain despite anatomic correction of ARAS.

2.1 Epidemiology

Atherosclerotic renal artery stenosis (ARAS) accounts for 90% of the lesions that impede blood flow within the renal artery. **ARAS typically involves the ostium and proximal 1/3 of the renal artery** and can also involve the adjacent aorta. Involvement of segmental renal arteries and higher order branches is uncommon. As atherosclerosis is a systemic condition, ARAS most often occurs in patients with risk factors for atherosclerotic disease, such as hypertension, hyperlipidemia, diabetes mellitus and tobacco use. Moreover ARAS is frequently identified in patients with cerebrovascular, coronary and peripheral arterial disease. **The prevalence of ARAS varies according to the radiographic definition (percent of luminal narrowing), imaging technique utilized and the patient population studied.** The prevalence in a general population of patients > 65 years of age was noted to be 6.8%, when assessed with duplex Doppler ultrasonography (DUS).³ In a systematic review of studies using more sensitive imaging techniques and defining ARAS as ≥ 50% luminal narrowing, the prevalence of ARAS ranged between 10.5% in consecutive patients undergoing coronary angiography to 54.1% in patients with congestive heart failure.⁹ In patients with underlying atherosclerotic conditions, ARAS can be anticipated in approximately 10-20% of cases. With these imaging techniques the prevalence of bilateral ARAS is 4.2%, in contrast to 12% when identified by DUS.^{3,9}

2.2 Pathophysiology

2.2.1 Hypertension

The physiologic basis for hypertension resulting from impaired renal blood flow was first elegantly described over 80 years ago.¹⁰ These initial studies stimulated numerous experiments that outlined the hormonal basis for the kidney's role in blood pressure control. From this work, it is well accepted that a reduction in renal perfusion leads to release of renin from the juxtaglomerular apparatus cells.

Renin, in turn, cleaves angiotensinogen (produced by the liver) to produce angiotensin I, which is then converted by the angiotensin converting enzyme (ACE) in the pulmonary vasculature to angiotensin II (Ang II). In addition to causing hypertension via its potent vasoconstrictive effects, Ang II stimulates adrenal synthesis of aldosterone, which further results in sodium and fluid retention.

In the classic 2-kidney, 1-clip model, increased renin release from the affected kidney leads to hypertension and sodium retention. However, the contralateral (unaffected) kidney senses increased perfusion pressure which, in turn, promotes a natriuresis and volume loss. **Therefore maintenance of hypertension in unilateral renal artery stenosis and normal contralateral kidney is primarily driven by renin and angiotensin. In contrast, in the setting of bilateral renal artery stenosis or renal artery stenosis affecting a solitary kidney, sodium and volume retention are not modified to the extent of having a normal contralateral kidney. Renin and angiotensin levels do not remain elevated and hypertension remains sodium and volume dependent**

Given the transient elevation in renin and angiotensin, several other pressor pathways participate in the development of hypertension. These pathways include sympatho-adrenergic activation, oxidative stress pathways and impaired vasodilatory responses within the kidney and systemic microcirculation.¹¹

2.2.2 Ischemic Nephropathy

A reduction in renal perfusion from ARAS that leads to renal parenchymal loss and declining renal function describes a condition known as ischemic nephropathy.¹² Experimental models suggest that impaired blood flow and an atherosclerotic milieu amplify tissue fibrosis and blunt remodeling pathways.¹³ Increased expression of pro-fibrotic mediators including transforming growth factor β (TGF-β) has been noted in these models. More recent studies have also identified renal microvascular changes distal to renal artery stenosis.¹⁴ Following an early period of microvascular proliferation, the continuing fibrotic process leads to loss of distal arterioles beyond the stenosis, further impairing renal function. Although presently there is no radiographic study that proves the diagnosis of ischemic nephropathy, the finding of disparity in kidney size on US, CT or nuclear medicine examination in combination with hypertension are strongly suggestive of ischemic nephropathy.

2.3 Diagnostic Evaluation

The primary goal when undertaking diagnostic evaluation for suspected RAS is to identify patients who, if found to have a lesion, are most likely to benefit from treatment. The decision to pursue diagnostic assessment, particularly with invasive studies, should be driven by the probability that renovascular disease is a significant contributor to the patient's condition. If blood pressure and renal function are satisfactorily controlled with medical therapy, further diagnostic evaluation may not lead to treatment gains.

In general, the clinical criteria that should prompt evaluation are (i) **early onset of hypertension** (age < 30 years), (ii) **accelerated hypertension**, (iii) **severe hypertension**, or (iv) **hypertension**

resistant to optimal medical therapy, (v) asymmetric kidneys with > 1.5 cm difference (vi) unexplained loss of kidney function (vii) rapid or recurrent decline in GFR in association with systemic blood pressure reduction, and (viii) decline in GFR (serum creatinine (Scr) \geq 30% from pre-treatment level) after initiation of ACE inhibitors or angiotensin receptor blockers (Table 3). Estimated GFR is now recommended to be independent of racial and ethnic background, calculated based upon patient age, gender, and either serum creatinine or cystatin-C concentration.¹⁵ A constellation of symptoms distinct to renal artery stenosis including acute flash pulmonary edema, unstable angina as a result of rapidly rising hypertension or recurrent congestive heart failure in a patient with adequate left ventricular function should raise suspicion of defective natriuresis. ¹⁶ **The guiding principle for diagnostic evaluation in ARAS is to evaluate patients who do not improve or who decline while on medical therapy.**

Table 3. Clinical criteria suggestive of atherosclerotic renal artery stenosis

Onset of hypertension at young age (< 30 years)
Accelerated hypertension
Severe hypertension
Hypertension resistant to optimal medical therapy
Asymmetric kidneys with > 1.5 cm length difference
Unexplained loss of kidney function
Rapid/recurrent decline in GFR associated with blood pressure reduction
≥30% decline in GFR after initiation of ACEIs or ARBs

2.3.1 Radiographic Studies

Radiographic evaluation of suspected ARAS should begin with non-invasive imaging studies. **Given the wide availability, low cost and ease to the patient, DUS is an excellent initial imaging study.** DUS can provide anatomical information relative to the renal parenchyma (kidney size, parenchymal thickness and echogenicity) as well as arterial blood flow characteristics within the main, accessory and intraparenchymal renal arteries.¹⁷ These blood flow characteristics include peak systolic velocity, end diastolic velocity, and adjacent aortic blood flow velocity. The ratio of velocity in the main renal artery to the aorta and resistive index ($RI = [(PSV-EDV)/PSV]$) can be derived from these measurements. In a recent study,¹⁸ $PSV > 200$ cm/s resulted in a sensitivity of 97%, specificity of 72%, PPV of 81% and NPV of 95% in terms of diagnostic accuracy for RAS. DUS has an added advantage that it can be performed serially for radiographic surveillance.¹⁸ The primary limitations of DUS are its high operator dependency and poor image quality in patients with large body habitus.

Computed tomography (CT) and magnetic resonance imaging (MRI) have sensitivity over 90% in detecting ARAS and MRI has a specificity of 91%.¹⁹ Both CT and MRI provide excellent anatomic detail however the blood flow changes that result from the stenosis cannot be determined. The functional consequences of these lesions may be indirectly determined by the findings of parenchymal atrophy. Another drawback of CT is the cost and need for intravenously-administered radiographic contrast agents. As many patients with suspected ARAS may have co-existent renal insufficiency, administration of iodinated contrast agents raises concerns of contrast nephropathy. Patients with $Scr \geq 1.5$ mg/dl should receive intravenous hydration before contrast administration and those with $Scr \geq 2.0$ mg/dl should not receive iodinated contrast as part of a diagnostic procedure (contrast should be reserved for use during a therapeutic procedure). Administration of intravenous gadolinium for MRA in patients with $GFR < 30$ ml/min should be avoided as it is associated with the development of nephrogenic systemic fibrosis. In these patients, it may be possible to use ferumoxytol (off label) as a blood pool contrast agent with MRI.²⁰

Renal arteriography, the “gold standard” for the diagnosis of ARAS, can also provide detailed imaging of the aorta, segmental renal arteries and intrarenal vasculature. Digital subtraction improves resolution and allows a reduction in the amount of contrast material. Angiography is an invasive study with risks of bleeding from arterial puncture, arterial dissection, arterial spasm and thromboembolism. Additionally iodinated contrast material cannot be administered in patients with contrast allergy or significant renal dysfunction and use of carbon dioxide may reduce the amount of contrast.

The diagnosis of ARAS is suspected by the finding of significant (> 50%) narrowing of the luminal diameter of the renal artery. As pressure differences across an arterial stenosis are exponentially related to diameter reduction, marked changes in pressure do not occur until the diameter reaches a critical threshold (approximately 70% of original diameter).²¹ The importance of this concept is that lower degrees of renal artery luminal narrowing are unlikely to impact kidney perfusion. Once the vessel diameter reaches this critical level, RAS becomes hemodynamically significant as evidenced

by a >10% drop in blood pressure across the stenosis.²²

Two additional diagnostic studies that warrant discussion are **radionuclide renography** and **plasma renin determination**. With radionuclide or nuclear renography, a radiopharmaceutical agent, either diethylenetriaminepentaacetic acid (DTPA) or mercaptoacetyltriglycine (MAG-3), is administered intravenously and images are reviewed to determine a renal functional difference, delay in time to peak uptake of the radioisotope or delayed excretion of the isotope. Additional imaging after administration of captopril, an ACE inhibitor, may show decreased function (GFR), suggesting the influence of angiotensin II maintaining glomerular filtration. Radionuclide renography has largely fallen out of favor due to its unreliability. In addition to diagnostic limitations in patients with diminished renal function ($\text{Scr} > 2.0 \text{ mg/dl}$) or bilateral arterial disease, these imaging studies have poor sensitivity and specificity.²³ Currently the primary role of renography in evaluating patients with hypertension may be to determine differential function prior to recommending nephrectomy.

Plasma renin activity or determination of renin level from the renal vein has been suggested to be highly diagnostic for the presence of renal artery stenosis but these studies are limited by their invasiveness and stringent testing conditions. The patient's volume status must be optimized, concurrent medications must be addressed and blood pressure tightly controlled. Lastly, and as mentioned previously, renin may be elevated early in the course of ARAS and likely declines over time as other pathophysiologic processes maintain the hypertensive state.

2.4 Treatment Options for ARAS

2.4.1 Medical Management

Medical management comprises the foundation of ARAS treatment. The general objectives of therapy are control of blood pressure, preservation of renal function and prevention of atherosclerosis-related complications. Antihypertensive treatment with ACE inhibitors or ARBs are often a first-line approach, both for blood pressure control and for their beneficial effect on cardiovascular risk reduction.²⁴ Use of these agents may initiate concerns of renal insufficiency from loss of glomerular filtration as a result of blockade of the Ang-II-induced efferent arteriolar constriction. However, significant renal function decline is uncommon in most series, and when it occurs, is virtually diagnostic of renal artery stenosis. If additional anti-hypertensive agents are needed, beta-blockers, diuretics and calcium-channel blockers can be added.

Given the presence of systemic atherosclerosis, many patients with ARAS have pre-existing cardiovascular risk factors including essential hypertension, diabetes mellitus, dyslipidemia and smoking. Established adjunctive therapies include glycemic control, statins, anti-platelet agents and lifestyle modifications including tobacco cessation.

2.4.2 Procedural Management

Whether all patients with ARAS should undergo renal revascularization in addition to medical therapy remains a controversial practice although recent trials do not support this procedure. Renal revascularization, both with open surgical renal artery bypass and percutaneous transluminal

angioplasty ± stent placement, has been the subject of several randomized prospective trials. Numerous single center retrospective studies have reported on various techniques of open surgical revascularization and have shown durable long-term patency rates.^{25,26,27,28,29,30,31} Notwithstanding these results, open surgical renal artery repair is associated with considerable perioperative mortality (5%) and morbidity (20%) rates, and therefore is generally reserved for young patients and those with hemodynamically significant ARAS and significant juxtarenal or suprarenal aortic disease.³²

Renal revascularization with percutaneous transluminal renal arterioplasty (PTRA) has been compared to medical therapy in three randomized controlled trials, none of which showed improved renal function, hypertension management or survival with PTRA.^{33,34,35} These trials enrolled small numbers of patients, excluded patients with severe renal functional impairment, had extensive crossover between treatment groups and did not utilize current medical therapy; thus making the results difficult to apply to current practice. Patients presenting with flash pulmonary edema are a high risk group and had a significant reduction in mortality and cardiovascular events with revascularization compared to medical management alone.³⁶

When compared to PTRA, PTR stenting (PTRS) was found to have superior technical success rates, increased patency rates and decreased restenosis rates leading to widespread adoption of PTRS as the standard endovascular management for ARAS during the 1990s and early 2000s.^{37,38} A comparison of PTRS and medical management for ARAS has been the subject of several randomized controlled trials. In three of these trials, which were presented or published in 2009, nearly 1000 patients with ARAS were enrolled and followed for 2-4 years.^{39,40,41} Aside from a slight reduction in number of anti-hypertensive medications, no significant improvement in systolic or diastolic blood pressure, renal function decline or adverse cardiovascular events was seen with PTRS in comparison to medical management. Despite the randomized prospective design, these trials have faced several criticisms including discrepancies in ARAS diagnosis, flaws in the intention-to-treat analysis and inclusion of patients with wide ranging ARAS. The recently published, NIH sponsored Cardiovascular Outcomes in Renal Atherosclerotic Lesions, or CORAL, trial attempted to correct these limitations and definitively answer whether revascularization of hemodynamically significant ARAS in hypertensive patients prevents adverse cardiovascular and renal events when added to optimal medical therapy.⁴² **The CORAL study enrolled 947 patients with ARAS and hypertension between 2005 and 2010 and randomly assigned these patients to medical therapy plus renal artery stenting or medical therapy alone. Following a median follow-up period of 43 months, the rate of the primary composite end point, comprised of cardiovascular and renal sequelae, was not significantly different between participants who underwent stenting in addition to medical therapy versus those who received medical therapy alone.**⁴² Although the limitations of these studies continue to be debated, the conclusions reached by the CORAL study reliably show the lack of benefit of renal artery stenting in the majority of patients with ARAS.

Complications of PTRS, although generally less frequent than those seen with open surgical revascularization, can still account for significant morbidity. Peri-procedural mortality is approximately

0.7% and renal artery complications range from 1 to 10%.⁴¹

3. Fibrous Dysplasia

Fibromuscular dysplasia (FMD) is a non-atherosclerotic, non-inflammatory vascular disease that most often affects small to medium arteries. Unlike atherosclerosis, FMD mostly involves the **mid to distal renal artery** and can affect segmental renal artery branches. **FMD has a female predominance and typically presents in patients between 20-60 years of age** but can occur in older and pediatric patient populations. Asymptomatic patients are most often diagnosed on imaging studies. Symptomatic patients can present with sequelae of the arterial disease such as stenosis, occlusion, arterial dissection or aneurysm. Although FMD can occur in any arterial bed in the body, involvement of the renal artery is most common and will be the focus of this chapter.

3.1 Epidemiology

Since many patients with FMD remain asymptomatic, the true prevalence of FMD is difficult to assess. The overall prevalence can be estimated from the evaluation of renal donors, a suitable representation of healthy adult population. The prevalence of FMD in renal donors ranges between 2.6-6.6% with the variance most likely due to differences in imaging technique and patient populations.^{43,44,45} Bilateral renal artery involvement is seen in approximately one-third of FMD cases.^{45,46} FMD has a marked female preponderance as nearly **90% of cases occur in women**. Several causative factors, including hormonal and environmental causes, have been proposed for the development of FMD, however the prevailing thought is that FMD is idiopathic.

3.2 Histopathology

FMD is classified according to the affected layer of arterial wall; specifically, the intima, media and adventitia (**Table 4**). FMD involving the media occurs most commonly and can be further subdivided into **medial fibroplasia**, **perimedial fibroplasia** and **medial hyperplasia**. The latter 2 conditions are very uncommon whereas **medial fibroplasia accounts for 90% of all types of FMD**. Medial fibroplasia is characterized by multiple diaphragm-like thickened areas of media that result in segments of arterial stenosis and poststenotic dilatation, typically seen as a “string of beads” appearance on angiography.⁴⁷ Accounting for the remaining 10% of cases of FMD, intimal fibroplasia is due to collagen deposition within the intima with resultant fragmentation of the intimal layer. On angiography, intimal fibroplasia often appears as a concentric stenosis and, occasionally as a long, diffuse narrowing.

Table 4. Fibromuscular dysplasias of the renal artery

Medial fibroplasia	~ 90%
Perimedial fibroplasia	rare
Medial hyperplasia	rare
Intimal fibroplasia	~ 10%

3.3 Clinical Presentation

As mentioned previously, the diagnosis of renal artery FMD is often made in asymptomatic patients undergoing abdominal imaging for other reasons. The most common symptomatic presentation is **a middle aged woman with new-onset or difficult to control hypertension**. Other clinical presentations of renal artery FMD include flank pain and/or hematuria, both of which can result from dissection or occlusion of the renal artery and aneurysmal changes involving the artery. Interestingly, unless dissection occurs, medial fibroplasia rarely results in renal dysfunction. Intimal and perimedial fibroplasia may be associated with renal dysfunction, arterial dissection and progression to occlusion.⁴⁸

3.4 Diagnostic Evaluation

As with ARAS, angiography remains the “gold standard” to diagnose and evaluate renal artery FMD. Catheter based angiography provides excellent resolution of the main and segmental renal arteries and can identify luminal changes such as aneurysm formation and dissection that may affect the smaller vessels. More recently intravascular ultrasound has been used as an adjunct to angiography and can depict pressure gradients and even provide virtual histology of the arterial lumen.⁴⁹ CT and MRA are able suggest the diagnosis of FMD in main renal vessels but may be limited by poor visualization along branch renal vessels.

3.5 Management of FMD

The primary treatment goal in patients with renal artery FMD is control of hypertension and prevention of secondary complications of hypertension. Since ischemic nephropathy is not a sequela of FMD, preservation of renal function is not a consideration in the treatment of FMD. The primary factors in considering which patients with FMD merit treatment are patient age, temporal relationship between onset of hypertension and diagnosis of FMD, adequacy of blood pressure control with anti-hypertensive agents and duration of hypertension.

Asymptomatic, normotensive patients with renal artery FMD should be followed for the development of hypertension. Young, otherwise healthy patients in whom FMD is diagnosed proximate to the onset of hypertension should be referred for PTCA, which provides immediate and long-term blood pressure control in nearly 75% of patients^{50,51} Patients in whom hypertension has been present for many years should be continued on anti-hypertensive medications as long as blood pressure control is satisfactory. **DUS-based surveillance of kidney length and cortical thickness should be done once or twice per year.**

If blood pressure control becomes difficult, medication side-effects become intolerable or renal size or function decrease, PTCA should be performed.⁴⁶ Balloon angioplasty alone is a very effective treatment for renal artery FMD thereby making stent placement unnecessary. Surgical revascularization is reserved for cases not amenable to percutaneous approaches such as those with aneurysms or FMD involving distal extrarenal branches. Indications for stent placement in FMD

are inability to eliminate a pressure gradient with angioplasty alone and intimal dissection.

Treatment success is fairly similar between PTRA and surgery. Approximately 50% of patients have cure of hypertension but the complications are markedly fewer following PTRA. Major complication rates following PTRA are approximately 12%, in contrast to 17% following surgery.⁵¹

Restenosis can occur after PTRA, particularly if the angioplasty was inadequate. Most instances of restenosis can be successfully treated with repeat angioplasty. Use of intravascular ultrasound (IVUS) in this setting can document resolution of a pressure gradient. Similar to patients on anti-hypertensive medications, patients are advised to continue with routine surveillance for recurrence of hypertension and for DUS of renal length and cortical thickness. Routine use of aspirin (81 mg) is advised for all patients with renal artery FMD.⁴⁶

4. Renal Artery Aneurysm

Renal artery aneurysm (RAA) is a dilation of the renal artery and/or branch arteries due to weakening of the elastic tissue and arterial media. In contrast to a false or “pseudo-aneurysm”, a true aneurysm involves all 3 layers of the arterial wall.

4.1 Epidemiology

The prevalence of RAAs is difficult to determine but can be estimated from several renal imaging analyses. In patients undergoing angiography, the prevalence ranges between 0.3-1.32%.^{52,53,54} As mentioned previously, patients with FMD appear to have a higher prevalence of RAAs. This notion is supported by recent studies of healthy kidney donors in which potential donors with radiographic findings of FMD had concomitant findings of RAA in 4.4-8.5% of cases.^{44,45}

4.2 Pathology

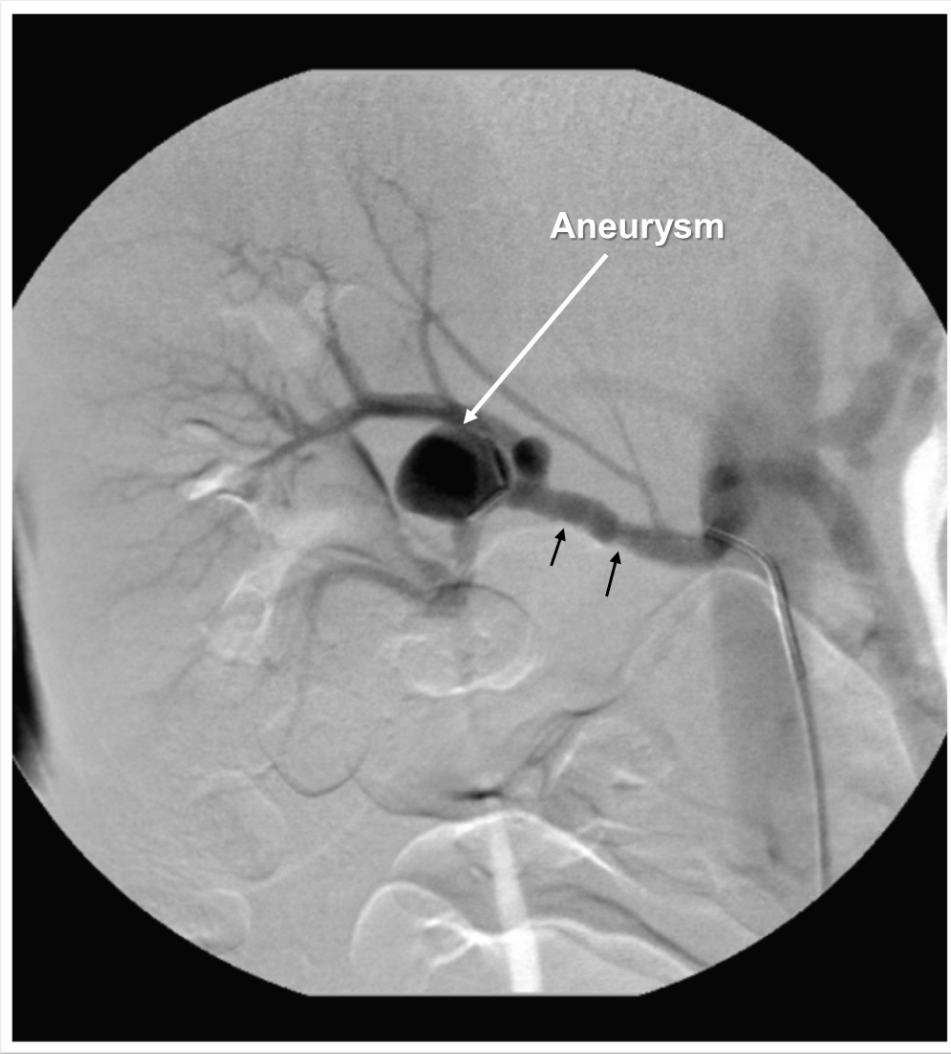


Figure 1: Saccular aneurysms at the bifurcation of the main renal artery.

RAAs can be classified as **saccular**, **fusiform**, **dissecting** and **intrarenal**.⁵⁵ Saccular aneurysms, the most common type, account for 70-90% and typically occur at the bifurcation of the main renal artery or distal branches (Figure 1). A uniform dilation of a renal artery segment occurs in fusiform aneurysms. These aneurysms may appear as areas of post-stenotic dilatation distal to an FMD lesion. In dissecting aneurysms, the intimal and portion of the medial layer disrupt and separate from the remainder of the arterial wall. The dissection can extend to the smaller branches and lead to limitation in flow, arterial thrombosis and infarction of the end organ. Intrarenal aneurysms are found within the renal parenchyma.

4.3 Diagnosis and Clinical Presentation

Most cases of RAA are identified on radiographic studies. In a previous era of plain-film radiography and intravenous pyelography, an “egg-shell” like pattern of calcification overlying the kidney was very suggestive of a RAA. With current imaging techniques, RAA are accurately diagnosed on CT scans. Using non-contrast scans, rim-like calcifications, particularly outside the renal parenchyma or

collecting system, should raise the suspicion for an aneurysm. The caliber and course of the renal artery can then be accurately assessed following administration of intravenous contrast. **The differential diagnosis of RAA includes enlarged and possibly calcified lymph nodes, calcified renal cysts and aneurysms of adjacent visceral arteries.** Renal artery aneurysms can also be identified on MRA however the degree of calcification cannot be determined. Similar to its utility in other renal arterial disease, renal arteriography is an excellent means for both the diagnosis and treatment of RAA.

Most patients in whom RAAs are diagnosed are asymptomatic at the time of diagnosis and remain without symptoms throughout follow up. Patients can present with hypertension and/or flank pain due to distal embolization and renal infarction. Hematuria, gross or microscopic, can also occur as a result of tissue infarction and from erosion of the aneurysm into the pelvicalyceal system. Auscultation of an abdominal or flank bruit may also indicate the presence of an aneurysm.

4.4 Management of RAA

In general, patients with symptoms or signs reliably attributable to the RAA should undergo treatment of the aneurysm. The difficulty arises in considering treatment of asymptomatic patients. **The fundamental judgment in managing asymptomatic patients is assessment of the risk of rupture.** Two prospective studies of patients with small, asymptomatic RAA suggest the risk of rupture to be very low.^{56,57} Clearly the risk of rupture in any aneurysm is related to size, specifically luminal diameter. **With RAAs the critical diameter for increased risk of rupture appears to be 2 cm . Additional risk factors for rupture are lack of calcification, severe, uncontrolled hypertension and potential for pregnancy (Table 4 and Table 5).** It is postulated that the increased cardiac output and hormonal influences on the vascular wall account for the relationship between pregnancy and risk of rupture. Based upon these criteria, treatment should be recommended for asymptomatic patients with one or more of these risk factors and for patients in whom there is marked increase in the luminal diameter.

Table 5. Indications for renal artery aneurysm repair

Asymptomatic patients	Symptomatic patients
Diameter ~ 2 cm and minimal to no calcification	Perinephric or retroperitoneal bleeding
Uncontrolled and/or multidrug resistant hypertension	Segmental or global renal ischemia/infarction
Woman of childbearing age with potential for pregnancy	Gross hematuria secondary to renal infarction or erosion into collecting system

Treatment of RAA comprises exclusion of blood flow from the aneurysmal portion of the artery. Open surgical repair has historically been very successful for aneurysm repair and renal salvage. Operative techniques for RAA repair vary according to the location of the aneurysm. Aneurysms along the main renal artery can be repaired by aneurysmectomy and primary repair of the artery or patch grafting. Vascular bypass to a target distal to the aneurysm can also successfully exclude the RAA. For complex aneurysms and those involving renal artery branches, ex-vivo reconstruction and renal autotransplantation is preferential (**Figures 2-4**).⁵⁸

Surgeons with exceptional robotic skills have performed intracorporeal repair as demonstrated by this elegant video.

More recently endovascular techniques are being increasingly applied to RAA repair. Selective embolization with microcoils or microparticles promotes thrombosis of the aneurysm. The main limitation for this approach is the feasibility of accessing the aneurysm “neck”, in order to position the thrombogenic particles selectively in the aneurysm and not to affect a significant portion of renal parenchyma. Technological advances have also allowed covered stents to be used for aneurysm exclusion.⁵⁹

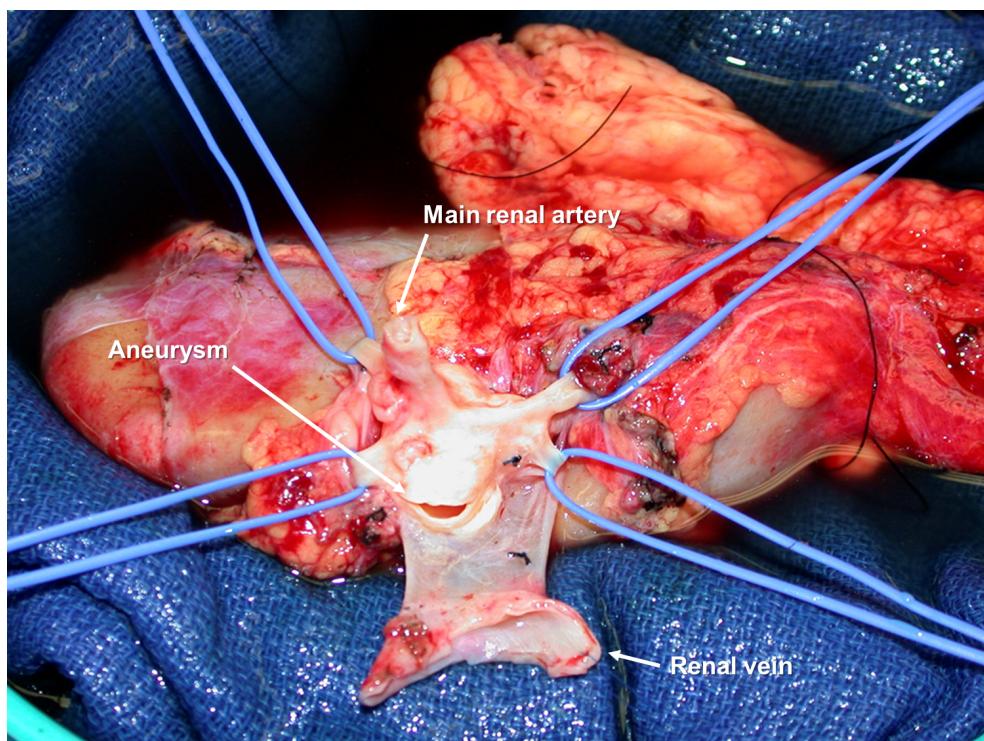


Figure 2

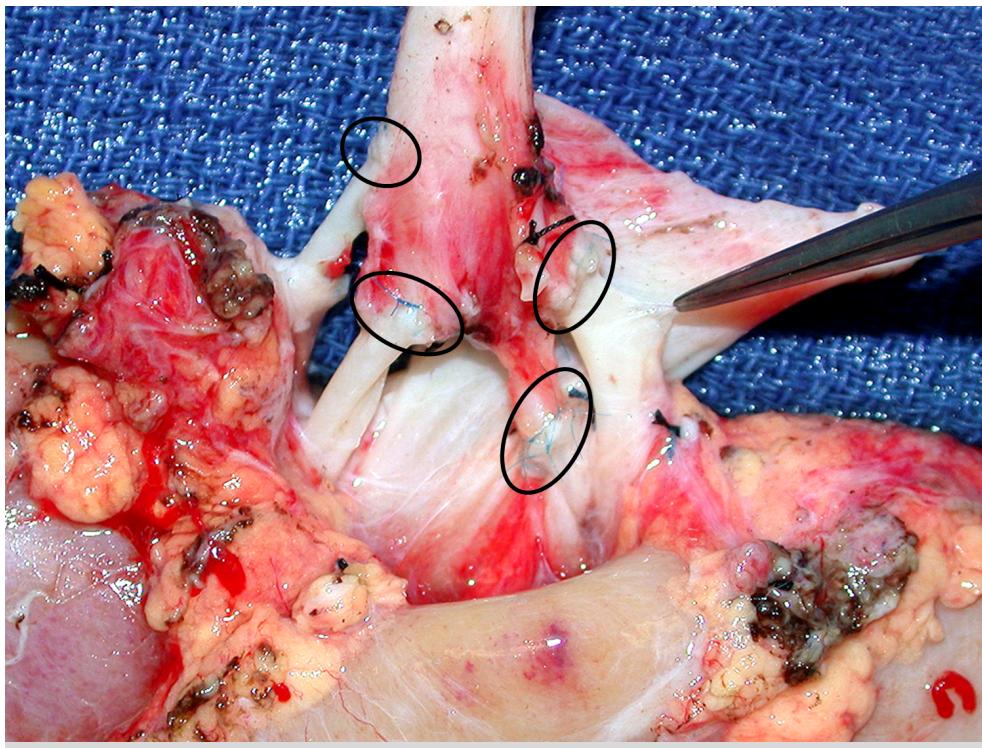


Figure 3

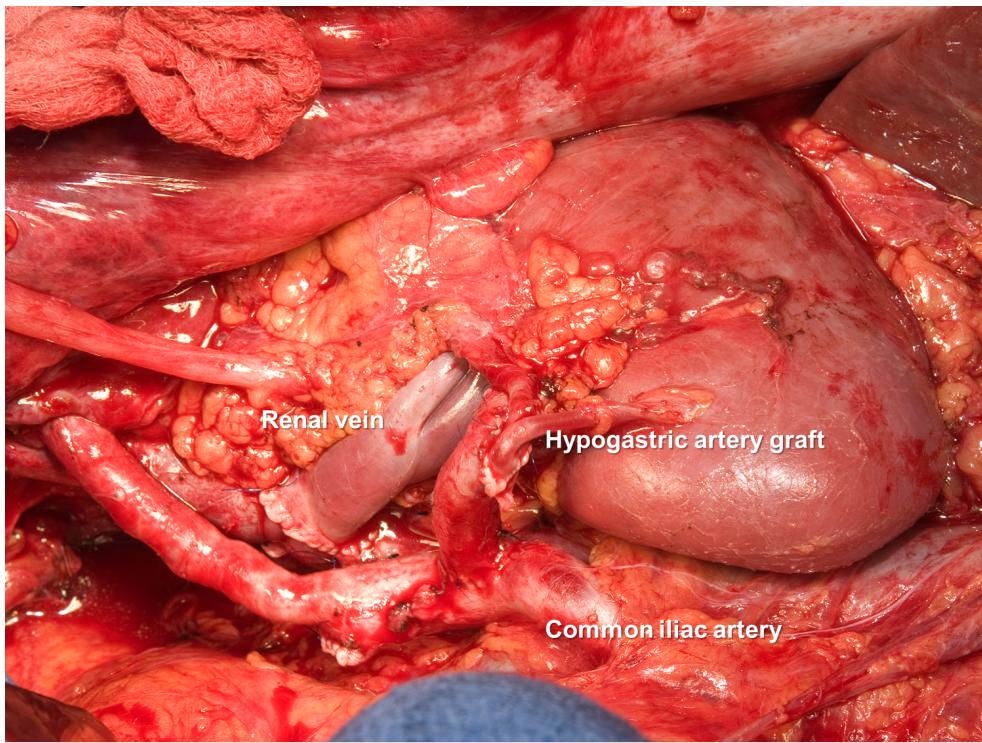


Figure 4

5. Abbreviations

ARAS: atherosclerotic renal artery stenosis

ACE: angiotensin converting enzyme

All: angiotensin II

ARB: angiotensin receptor blockers

CT: computed tomography

DUS: duplex ultrasonography

GFR: glomerular filtration rate

MRA: magnetic resonance angiography

PTRA: percutaneous transluminal renal artery angioplasty

PTRS: percutaneous transluminal renal artery stent placement

RAA: renal artery aneurysm

RAS: renal artery stenosis

sCr: serum creatinine

Videos

Robotic Renal Artery Aneurysm Repair with Intra-arterial Renal Hypothermic Perfusion

Presentations

Renovascular Diseases Presentation 1

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