

# Pediatric renovascular hypertension in Thailand: CT angiographic findings

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## Abstract

**Background** Renovascular disease is an uncommon but important cause of hypertension in children. When unrecognized and untreated, renovascular hypertension in children can have serious complications.

**Objective** To review the causes of renovascular hypertension and computed tomography angiographic (CTA) findings in children and adolescents.

**Materials and methods** Twenty-eight CTAs from January 2004 to March 2008 of 23 children and adolescents with hypertension were reviewed for the causes and CTA findings.

**Results** Nine of the 23 children (39%) had abnormal renal arteries with or without abnormal abdominal aortas. Four of these children had Takayasu arteritis, one had moyamoya disease, and one had median arcuate ligament syndrome. One with chronic pyelonephritis had severe stenosis of the proximal right renal artery. The other two children had renal artery stenosis with a nonspecific cause. One child with a normal abdominal aorta and renal arteries had a right suprarenal mass. On pathological examination a ganglioneuroma was found.

**Conclusion** CTA can help in diagnosis of renovascular hypertension in children and adolescents. Although CTA is not a screening modality, it is appropriate in some situations.

**Keywords** Renovascular hypertension · Renal artery stenosis · Multidetector CT · Children

## Introduction

Renovascular disease is an uncommon but important cause of hypertension in children. It is usually diagnosed after a long delay because blood pressure is not easily measured in small children, and in some practices blood pressure measurements are not routinely obtained [1]. When unrecognized and untreated, renovascular hypertension in children can have serious complications, including hemorrhagic stroke, hypertensive encephalopathy with impaired mental development, and failure to thrive. Poorly controlled hypertension can result in left ventricular hypertrophy and severe diastolic dysfunction [2]. Various diseases are associated with childhood renovascular hypertension such as fibromuscular dysplasia, neurofibromatosis type 1, and vasculitis [1–3]. Takayasu arteritis is the most common cause of renovascular disease in Asia and Africa, whereas in most western countries fibromuscular dysplasia or vascular lesions from neurofibromatosis are the most common causes [4]. In a study of children with renovascular hypertension in India, the cause of the disorder was Takayasu arteritis in 85% of cases [5].

The diagnosis of renovascular hypertension can be established with several different imaging modalities such as Doppler US, renal scintigraphy, CT angiography (CTA), and MR angiography. However, conventional angiography is still the gold standard. The purpose of this study is to review the causes of renovascular hypertension and CTA

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findings in children and adolescents at a large referral hospital in Thailand.

## Materials and methods

The study protocol was approved by our institute's ethics committee. From January 2004 to March 2008, a total of 28 CTA studies were performed on 23 children and adolescents with hypertension. There were seven girls and 16 boys. Their ages ranged from 1 month to 18 years, with a median age of 13 years. Three children had two CTA studies and one had three CTA studies. Nineteen children presented with a variety of complaints and histories, including headache, seizures, dyspnea, anemia, blurred vision, obesity, and moyamoya disease. The other four children were found to have hypertension during physical examination for other reasons such as trauma and upper respiratory tract infection. Twelve children had had Doppler renal US and three had had captopril renography prior to CTA. Conventional angiography was done for five children.

Twenty-six CTA studies were done using a 16-slice multidetector CT (Aquilion 16, Toshiba, Japan) with the following parameters: 1-mm collimation, 15 mm per rotation table speed, and 0.5-mm increment. Two CTA studies were done using a dual-source CT scanner (SOMATOM Definition, Siemens Medical Solutions, Forchheim, Germany) by using 0.6-mm collimation, 12 mm per rotation table speed, and 0.4-mm increment. Most of the children were scanned using 120 kV and 175 mA. Children younger than 8 years were sedated by a pediatrician. CTAs in children younger than 8 years were performed during quiet breathing; CTAs in children 8 years or older were performed with breath-holding. A nonionic contrast medium was injected into the children's antecubital veins using a power injector, 300 mg iodine per ml with a dosage of 2 ml per kilogram. The injection rate was 2–2.5 mL/s. Scans were obtained from the dome of the diaphragm to the aortic bifurcation at the aortic peak of the time-density curve. All the data were then transferred to a workstation using Voxar 3D<sup>TM</sup> software (Voxar Inc., Edinburgh, UK). For each CT examination, axial images supplemented by 2- and 3-dimensional postprocessing including multiplanar reformations, maximum-intensity projections, and volume-rendering techniques were used to review for abnormalities of the abdominal aorta and renal arteries. The grading of renal stenosis was as follows: mild (less than 50% stenosis), moderate (50–75% stenosis), severe (more than 75% stenosis), and occlusion. Medical records of these children were also reviewed for the diagnoses. The diagnoses of children with CTA findings of renal artery stenosis were confirmed by conventional angiography, surgery, or clinical

history. Comparison of CTA with conventional angiography findings was not done because the time between the two studies varied from 3 months to 1 year and 5 months.

## Results

CTAs of 14 of the 23 children (61%) revealed normal abdominal aortas and normal renal arteries. One of these children had a right suprarenal mass. He underwent right adrenalectomy; on pathological examination a ganglioneuroma was found. Nine children (39%) had abnormal renal arteries with or without abnormal abdominal aortas; the details are in Table 1. For children who had more than one CT examination, the results of the last CT examinations are listed in Table 1. Four of these nine children had Takayasu arteritis, one had moyamoya disease, and one had median arcuate ligament syndrome. One child with severe stenosis of the proximal right renal artery and a small right kidney (case 6) underwent right nephrectomy; pathological examination revealed chronic pyelonephritis and thrombosis of the renal artery. Two children with renal artery stenosis with a nonspecific cause (case 2 and case 5) did not have string-of-beads appearances of the renal arteries, as has been reported for fibromuscular dysplasia [6, 7]. These two children also underwent conventional angiography, which did not reveal any fibromuscular dysplasia. The stenoses of their renal arteries also occurred in the proximal portions.

CTAs of three of the four children with Takayasu arteritis showed thickened walls of the abdominal aortas. Two of these children also had dilatation of the abdominal aortas. The stenosis or occlusion of the renal arteries in these four children was at the ostium or in the proximal portion (Fig. 1). The abnormalities of other branches of abdominal aorta are described in Table 1. All children with Takayasu arteritis had positive tuberculin skin tests.

Two children in the study had moyamoya disease. One had a normal aorta and normal renal arteries. The other had severe segmental stenosis of the proximal right renal artery, moderate focal stenosis of the proximal portion of the left renal artery (Fig. 2), and occlusion of the posterior division of the left renal artery. This child had been diagnosed with moyamoya disease when he was 3 years old.

The child with median arcuate ligament syndrome had severe stenosis of the celiac artery, mild ostial stenosis of the right renal artery with occlusion of the proximal anterior division with associated renal infarction, and kinking of the proximal segment of the left renal artery from compression by the left diaphragmatic crus (Fig. 3). The abdominal aorta of this child appeared normal.

All children with Takayasu arteritis received antituberculous drugs. One underwent angioplasty. One of the two children with nonspecific renal artery stenosis underwent

**Table 1** CT findings of renal arteries and abdominal aorta and other branches in children with renovascular hypertension

No.	Age/ Sex (year)	CT findings		Diagnosis
		Renal arteries	Abdominal aorta and other branches	
1	8 /F	RRA: Ostial occlusion with multiple collaterals. LRA: Severe stenosis. Small LK.	Thickened wall of aorta with calcification. Celiac artery occlusion and SMA ostial stenosis.	Takayasu arteritis
2	3/M	RRA: Smooth tapered stenosis of proximal anterior and posterior divisions with poor perfusion. LRA: Normal.	Normal	Nonspecific renal artery stenosis
3	18/F	RRA: Severe focal stenosis of proximal RRA and another mild stenotic lesion of proximal RRA. LRA: Occlusion. Small LK.	Aneurysmal dilatation of supra- and infrarenal aorta.	Takayasu arteritis
4	5/M	RRA: Severe segmental stenosis of proximal RRA. LRA: Moderate focal stenosis of proximal LRA. Short occlusion of posterior division.	Normal	Moyamoya disease
5	14/M	RRA: Mild focal stenosis of proximal RRA. LRA: Moderate focal stenosis of middle LRA with post stenotic dilatation.	Normal	Nonspecific renal artery stenosis
6	11/M	RRA: Severe stenosis of proximal RRA. Small RK. LRA: Normal.	Normal	Chronic pyelonephritis
7	10/M	RRA: Ostial stenosis. LRA: Ostial occlusion.	Thickened wall of aorta with fusiform dilatation of suprarenal aorta. Celiac and SMA: ostial occlusion with collateral from IMA.	Takayasu arteritis
8	9/F	RRA: Severe ostial stenosis of the inferior RRA. Small superior RRA. Small RK. LRA: Mild ostial stenosis.	Thickened wall of aorta with mild segmental narrowing of mid-infrarenal aorta. Celiac and SMA ostial stenosis.	Takayasu arteritis
9	18/M	RRA: Mild ostial stenosis with occlusion of proximal anterior division with associated renal infarction. LRA: Kinking of proximal segment from compression by left diaphragmatic crus.	Normal aorta. Celiac artery: severe stenosis from median arcuate ligament syndrome.	Renal artery entrapment

SMA superior mesenteric artery; IMA inferior mesenteric artery; RRA right renal artery; LRA left renal artery; RK right kidney; and LK left kidney

angioplasty. The parents of the child with moyamoya disease refused angioplasty. The child with renal artery entrapment underwent angioplasty of the left renal artery, but the procedure was not successful. The child was lost to follow-up.

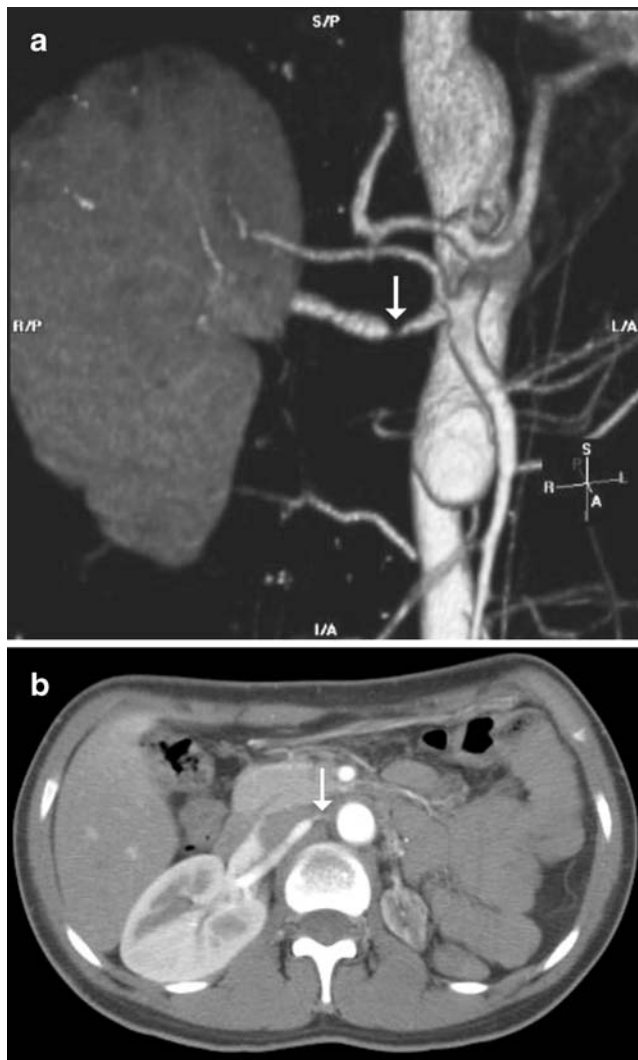
Of the 14 children with normal abdominal aorta and renal arteries, 6 had normal Doppler renal US, 1 had a normal value of plasma rennin activity, 2 had normal urine VMA tests, 1 had a negative I-131 MIBG scan, 2 had normal blood pressures at 1 month and 7 months after CTA, and 1 child was diagnosed as having metabolic syndrome. One child was lost to follow-up.

**Discussion**

Renovascular hypertension accounts for 5–10% of children with hypertension [5, 8]. In a survey of children age 17 and younger in Turkey, 21 children had bilateral renal artery stenosis (RAS) and 24 had unilateral RAS [8]. The most common cause was fibromuscular dysplasia. The other causes were Takayasu arteritis, neurofibromatosis, Williams syndrome, Kawasaki disease, mid-aortic syndrome, extrin-

sic compression of the renal artery, and nonspecific bilateral RAS [8]. In a study of malignant hypertension in Indian children age 16 and younger, Takayasu arteritis was found in 11 of 13 children with renovascular hypertension and fibromuscular dysplasia was found in 2 [5]. Takayasu arteritis is also a significant cause of renovascular hypertension in South African children [9]. In our study, the most common cause of renovascular hypertension was also Takayasu arteritis.

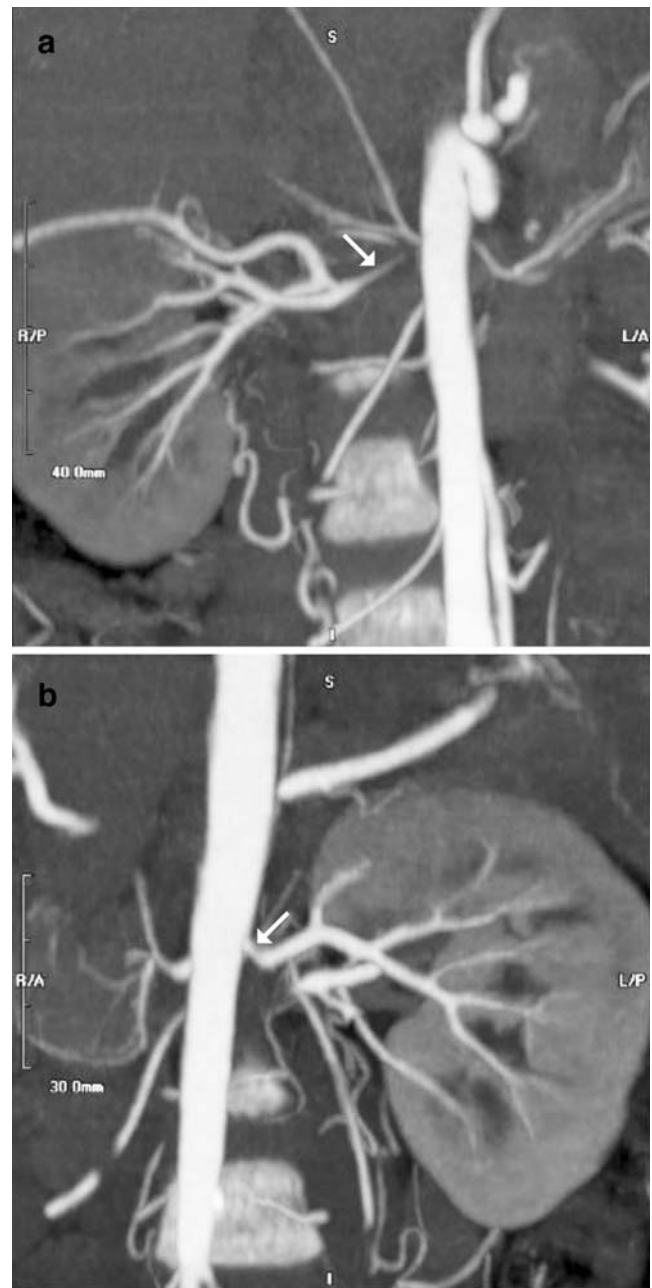
Takayasu arteritis is a chronic idiopathic inflammatory disease primarily affecting the aorta, its proximal branches, and the pulmonary arteries. It occurs in many parts of the world, but there seems to be a higher incidence in communities where tuberculosis is endemic [9]. Because the basic pathologic feature of early phase Takayasu arteritis is thickened walls of the great vessels, CT is useful for early diagnosis because it allows evaluation of wall thickness rather than merely the diameter of the lumen, which is especially important because early diagnosis and treatment improve prognosis. Moreover, CT scanning can provide information about wall enhancement during the active phase of disease [10]. The spectrum of findings on CTA includes stenoses; occlusions; aneurysms; and con-



**Fig. 1** Takayasu arteritis with renovascular hypertension. **a** CTA with maximum-intensity projection technique shows severe focal stenosis of the proximal right renal artery (*arrow*) and aneurysmal dilatation of the supra- and infrarenal aorta. **b** CTA axial image shows severe focal stenosis of the proximal right renal artery (*arrow*) and occlusion of the left renal artery with the small left kidney

centric arterial wall thickening affecting the aorta and its branches, the pulmonary arteries, and occasionally the coronary arteries. In the later stages of the disease, extensive vascular calcification can occur [11]. Renal involvement in Takayasu arteritis is usually caused by RAS. Renal angiography demonstrates stenosis or occlusion of the main renal artery resulting in renal infarcts in about 65% to 75% of patients [12].

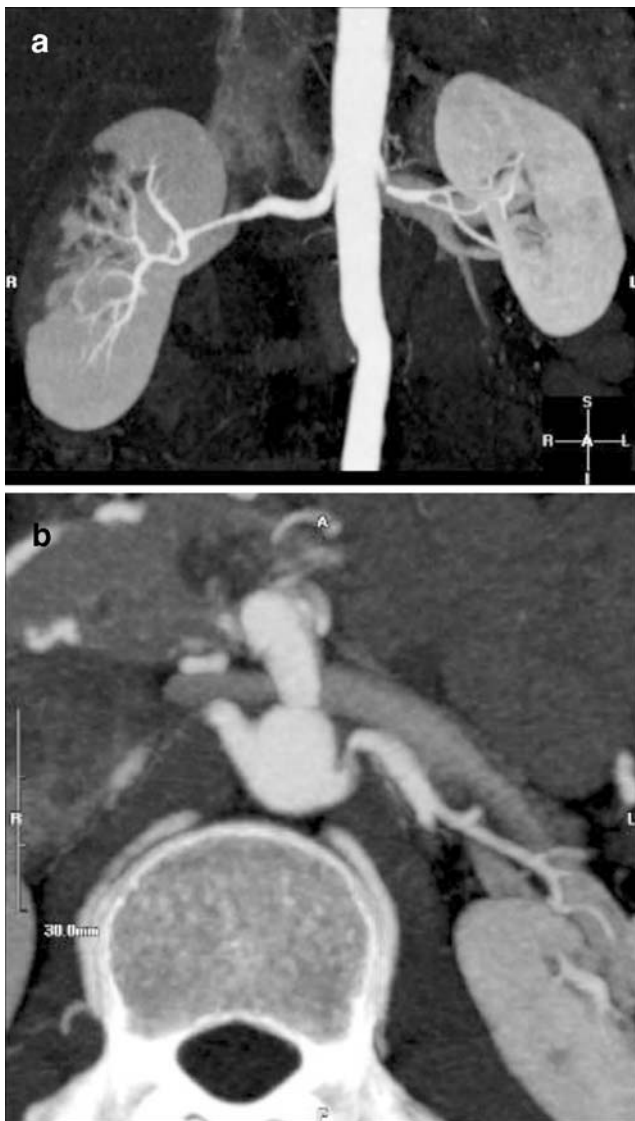
The etiology of Takayasu arteritis is unknown. However, tuberculosis might play a role in its pathogenesis or progression. The association between Takayasu arteritis and tuberculosis is variable. One study from South Africa reported that 92% of 26 children with Takayasu arteritis had



**Fig. 2** Moyamoya disease with stenoses of the proximal portion of bilateral renal arteries. **a, b** CTA with maximum-intensity projection technique shows severe segmental stenosis of the right renal artery (*arrow, a*) and moderate focal stenosis of the left renal artery (*arrow, b*). Note collateral vessels

tuberculosis [9], while none of 11 children in India had tuberculosis [5].

Moyamoya disease is a rare cerebrovascular occlusive disorder most often found in Japanese patients. It is characterized by stenosis or occlusion of the bilateral internal carotid arteries with abnormal vascular networks at the base of the brain. Vascular stenooclusive changes occur not only in the intracranial vessels but also in the extracranial ones [13]. Extracranial vascular involvement in



**Fig. 3** Renal entrapment in median arcuate artery syndrome. **a** CTA with volume-rendering technique and **b** CTA axial image show proximal portions of renal arteries pulled down and in toward the aorta, with kinking of the proximal left renal artery from left diaphragmatic crus compression. Note decreased enhancement of the lateral portion of the right kidney from infarction

moyamoya disease has been reported mainly in the renal artery, mostly in the proximal region of the main branch [13, 14]. The incidence of RAS in moyamoya disease has been reported as 5–8.5% [13–15]. RAS in these patients occurred unilaterally or bilaterally [13, 14]. Pathologic studies have found that obstructive lesions in the cerebral vessels were caused mainly by fibrous thickening of the intima and a small amount of lipid deposition [13]. The extracranial vessels may also have intimal fibrous thickening similar to that found in the intracranial vessels [13]. Our patient with moyamoya disease and renal artery stenosis

had stenosis of the proximal portions of both renal arteries. He also had a short occlusion of the posterior division of the left renal artery.

The median arcuate ligament (MAL) is a tendinous band that connects the medial borders of the diaphragmatic crura on either side of the aortic hiatus, posterior and superior to the origin of the celiac axis. The appearance of the ligament varies greatly, ranging from a well-defined ligamentous mass to an amorphous area of connective tissue. Low insertion or high origin of the celiac or renal artery relative to the MAL in some patients may cause extrinsic compression of the celiac, mesenteric, and renal arteries [16]. Thony et al. [17] have reported that renal artery entrapment should be suspected each time angiography shows a renal artery parallel to the aorta in the proximal part of its course and when an ostial concentric stenosis is discovered in a patient free of atheroma. Ilica et al. [16] have reported multidetector CT findings of patients with renal entrapment by MAL. The proximal portions of renal arteries were pulled down and in toward the aorta, with mild to moderate narrowing [16]. These findings were seen in our patient with median arcuate ligament syndrome.

CTA is a reliable, noninvasive procedure for the detection of RAS. For adults some authors have reported that the sensitivity and specificity of CTA in diagnosing significant RAS (more than 50% stenosis) was greater than 90% [18–20]. Secondary signs of RAS, such as poststenotic dilatation, decrease in renal size, cortical thinning, and decreased cortical enhancement, are also helpful in recognizing RAS on CTA [21].

The accuracy of CTA in diagnosis of renovascular hypertension in children is unknown. In a survey of renovascular hypertension in children in Turkey, only two of 45 children underwent CTA [8]. In a study of 24 children with suspected renovascular disease who underwent CTA, the authors did not report the sensitivity or specificity of CTA. However, they found a false-negative in a child with fibromuscular dysplasia [3]. In our study, the sensitivity and specificity could not be determined because only some of our patients had conventional angiography. Perhaps CTA has not been used very often because of technical factors: older scanners might have had limited spatial and contrast resolution of tiny pediatric renal arteries. Certainly, radiation dose is an additional consideration.

The parameters for CTA with dose reduction for children have recently been published [22, 23]. Compared with conventional angiography, CTA doses in children are competitive, at least from work done in cardiovascular assessment. The dose depends obviously on the length of the angiographic procedure [23]. By using scanner-generated CT dose index volume, the CT dose of a 13-year-old boy in this study was 6.6 mGy at 115 mAs and

120 kVp. This is lower than that reported for adults, 17 mGy at 120 kVp [24].

The limitations of this study are the small number of patients and lack of conventional angiography, the gold standard for comparison. At present CTA should not be used to screen for RAS because the site of stenosis in children is frequently in second order vessels [25], CTA might not be able to evaluate these vessels because of their small caliber and thus might miss these lesions. The involvement of second order vessels and high radiation dose of CTA makes angiography the preferred option. Using angiography, one can also treat RAS at the same time.

However, CTA is a promising method for detection of lesions of the larger renal arteries, and if future technology permits the reliable depiction of stenoses of smaller branches, it could even replace diagnostic angiography. CTA has been reported to be helpful in patients with Williams syndrome, neurofibromatosis, and Takayasu arteritis [26].

## Conclusion

Our study shows that CTA can help in diagnosis of renovascular hypertension in children and adolescents. Although CTA is not a screening modality, it is appropriate in some situations.

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