# Agenesis of infrarenal abdominal aorta

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## ABSTRACT

**Background:** Infrarenal aortic agenesis is an extremely rare condition that has been described in a few case reports in the literature. Imaging clues to distinguish infrarenal abdominal aorta agenesis from other abdominal aortic occlusions include: the absence of an aorta at the infrarenal level without peri-aortic soft tissue or thickening of the aortic wall that suggests vasculitis or mass, and collaterals, such as a Riolan arch, for the anomaly to be life reconcilable.

**Case Presentation:** In this case report, a 60-year-old female patient with a diagnosis of infrarenal segmental aorta agenesis is described. Computed Tomography (CT) angiography was performed to evaluate the vascular involvement of the patient who is followed up with sarcoidosis. It was observed that the abdominal aorta terminated abruptly at the level of the renal arteries, and refilled via collaterals 2 cm distal to this level.

**Conclusion:** Recognition of this very rare entity by the radiologist is important in guiding the clinician in terms of treatment follow-up protocol. It is aimed to make this entity easier to recognize by radiologists with this case report.

**Keywords:** Abdominal aorta agenesis, volume-rendered computed tomography, Riolan arch, collaterals.

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## Background

Although hypoplasia and coarctation of the abdominal aorta have been widely described in the literature, only a few case reports have been reported for abdominal aortic agenesis [1-3]. Mesenteric ischemia, renal failure, uncontrolled hypertension, and claudication are common clinical presentation forms of this entity. Clinical findings vary according to which segment is affected and the presence of collateral circulations. Definitive diagnosis can be made by catheter angiography, MR angiography, or CT angiography.

## Case Presentation

A 60-year-old female patient was admitted to the cardiology department 1 year ago with complaints of chest pain and headache. Her initial examination was normal including blood pressure and pulses. Her ambulatory blood pressure was 113/67 mm Hg. The blood sample was normal including Blood Urea Nitrogen (BUN) and creatinine. The treadmill test and echocardiography were normal. Because of the known history of familial Mediterranean fever and sarcoidosis, CT angiography was obtained to evaluate vascular involvement.

In CT angiography at the L1 level, the abdominal aorta terminated as renal arteries, and approximately 2 cm distal to this about L2 level it refilled via collaterals (Figure1). The abdominal aorta showed refilling just proximal to the bifurcation level through the collaterals between the Superior Mesenteric Artery (SMA) and Inferior Mesenteric Artery (IMA) known as the arch of Riolan. Both SMA and IMA were ecstatic. In addition, the lumbar arteries were forming collaterals with the aorta at the defined level (Figure 2). There was no aortic wall thickening or peri-aortic soft tissue which suggests vasculitis. Except for the defined level, the aorta, visceral, and peripheral branches were in normal calibration.

## Discussion

While hypoplasia describes longer segmental stenosis, coarctation is defined as local segmental stenosis. Besides agenesis is defined as the complete absence of aortic tissue.

Even though the etiology is still unclear yet, aortic stenosis may be caused by atherosclerotic disease, vasculitis (such as Takayasu arteritis), infection (congenital rubella), external compression (peri-aortic fibrosis, retroperitoneal tumors), and even hereditary syndrome (neurofibromatosis, Williams–Beuren Syndrome, fibromuscular dysplasia [4,5].

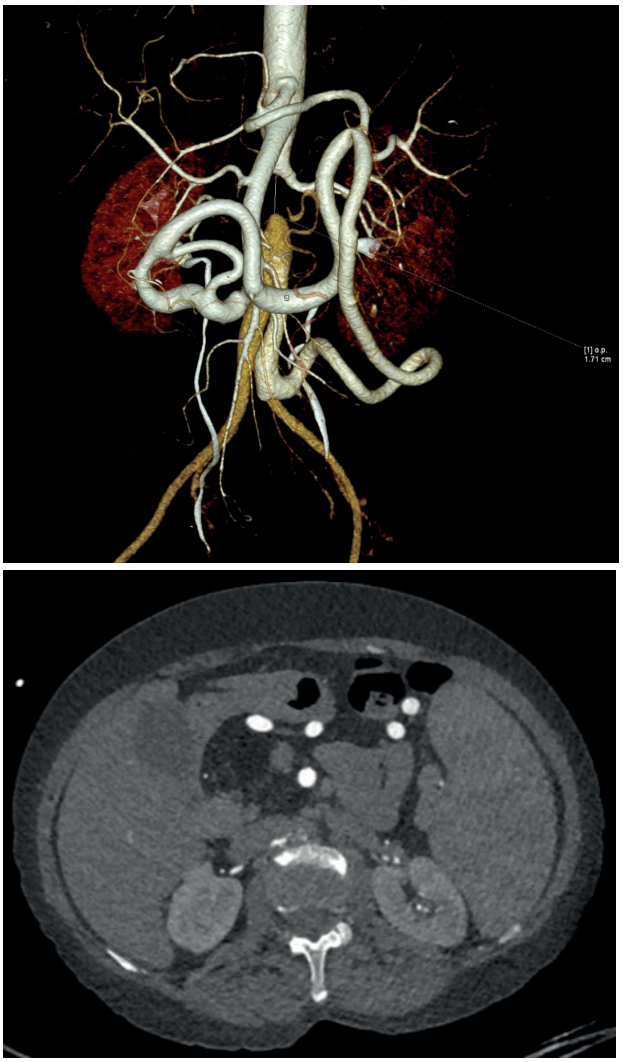

***Figure 1.*** *(a) Coronal 3D reconstructed volume rendered image demonstrates that the abdominal aorta shows abrupt termination at renal artery levels. Both renal arteries are normal calibration. The bifurcation level is normal. (b) Axial maximum intensity projection CT angiography image demonstrates the complete absence of aortic tissue. Only adipose tissue and lumbar collaterals are seen at that defined level.*

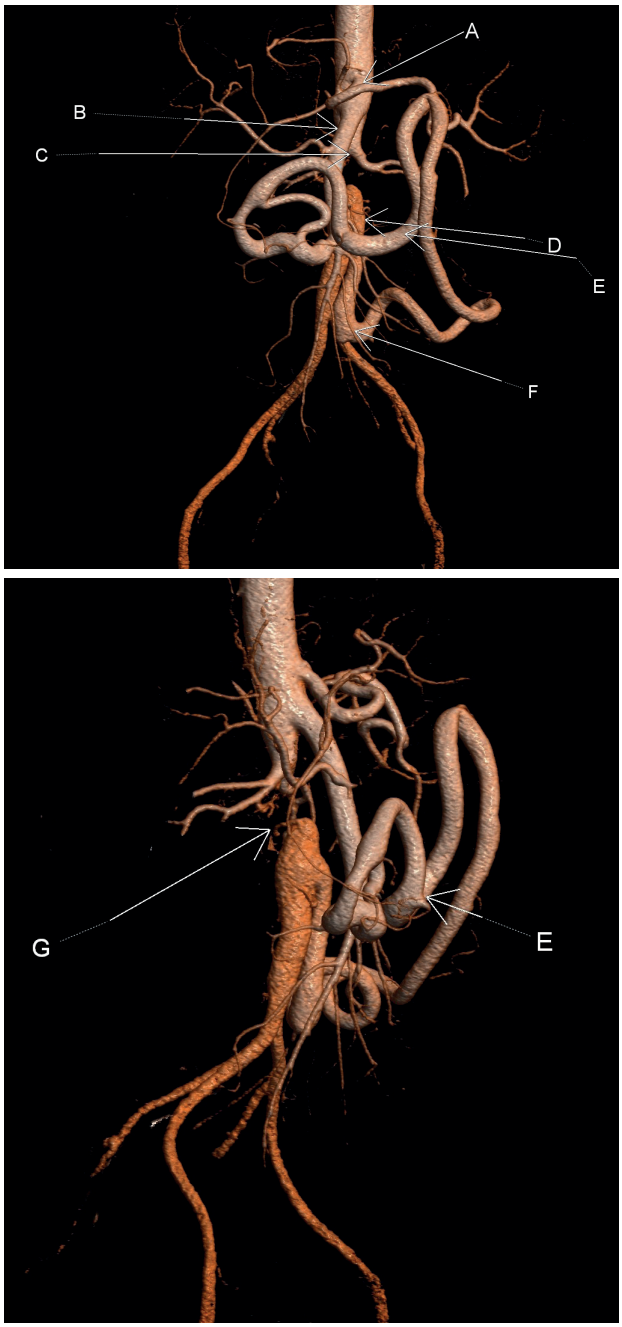


***Figure 2.*** *(a). Coronal 3D reconstructed volume rendered image demonstrates collateral vascular structures. A: Celiac trunk; B: SMA; C: Aorta termination level; D: Aortic bifurcation level; E: Arch of Riolan which connects SMA and IMA; F: IMA. (b). Sagittal 3D reconstructed volume rendered image demonstrates collateral vascular structures. E: Arch of Riolan which connects SMA and IMA; G: Lumbar collaterals.*

Congenital abdominal aortic stenosis is a rare anomaly and accounts for less than 2% of aortic coarctations [6]. Although coarctation and hypoplasia of the abdominal aorta are rare conditions, abdominal aortic agenesis has only been described in a few cases in the literature.

Diagnosis of abdominal aortic agenesis is often difficult due to its rareness and asymptomatic presentation. It is usually diagnosed incidentally, as in our case. The diagnosis is made by exclusion of extrinsic causes of stenosis (vasculitis, mass, etc.) and the absence of a certain level of aortic tissue [1,2].

Catheter angiography, CT angiography, or MR angiography are required for the definitive diagnosis. CT and MR angiography have the advantage of being easily accessible, fast, non-invasive, and producing 2D and 3D high-resolution reformatted images. The presence of affected segments and collateral structures determines visceral organ ischemia and extremity findings, and these findings determine the treatment or follow-up scheme.

## Conclusion

Recognition of this very rare entity by the radiologist is important in guiding the clinician in terms of treatment follow-up protocol. It is aimed to make this entity easier to recognize by radiologists with this case report. When radiologists encounter this disease, they should first be

able to distinguish between agenesis, hypoplasia, and coarctation, then the length of the absent segment in the abdominal aorta, collateral vascular structures, extremity, and visceral organ perfusion defects (if any) should be defined in detail.

### What is new?

In the literature, only a few case reports have been reported for abdominal aortic agenesis. Therefore, it is aimed to make this entity easier to recognize by radiologists with this case report.

### List of Abbreviations

| | |
|---|---|
| BUN | Blood Urea Nitrogen |
| CT | Computed Tomography |
| IMA | Inferior Mesenteric Artery |
| MRI | Magnetic Resonance Imaging |
| SMA | Superior Mesenteric Artery |

### Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

### Funding

None.

### Consent for publication

Informed consent was obtained from the patient to publish this case in a medical journal.

### Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

### Author details

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### Summary of the case

| | | |
|---|---|---|
| 1 | **Patient (gender, age)** | 60, female |
| 2 | **Final diagnosis** | Agenesis of infrarenal abdominal aorta |
| 3 | **Symptoms** | Chest pain and headache |
| 4 | **Medications** | N/A |
| 5 | **Clinical procedure** | N/A |
| 6 | **Specialty** | Radiology |