Mid-Aortic syndrome (MAS) - A rare but important entity!

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Learning objectives

• To review Mid-Aortic syndrome (MAS) clinical aspects and its correlation with imaging features.

• To illustrate key radiological findings of this syndrome.
Background

Mid-Aortic syndrome (MAS) is a rare entity characterized by narrowing of the abdominal aorta and its major branches. There will be involvement of the renal arteries in most patients. Although stenosis of the thoracic aorta is possible, it is uncommon. Typically affects children and young adults.

Most cases are idiopathic. There are several theories about its possible origin being the most accepted a probable lack of fusion of both dorsal aortas in embryonic life with atrophy and stenosis of one of them. MAS may also be secondary to other diseases such as Takayasu arteritis, Williams syndrome, mucolipidosis and neurofibromatosis. However, morphological findings are similar regardless of the etiology.

Hypertension is the most common symptom. The course of the disease depends on the degree of stenosis, the affected arteries and the progressive damage caused by renovascular hypertension. If left untreated, patients usually die due to renal failure or severe ischemic complications.
Findings and procedure details

Mid-Aortic syndrome (MAS) is characterized by stenosis of the abdominal aorta frequently associated with a concomitant stenosis of the renal and visceral arteries Fig.1. Artery stenosis typically involves the ostium of the vessel.

MAS is typical of childhood without a predilection for sex. In most cases it is idiopathic. Although there is no clear consensus on the mechanism by which it occurs, a congenital anomaly has been suggested. A lack of fusion of both dorsal aortas in embryonic life is the most accepted theory. What is clear is that it can be secondary to several genetic syndromes with vascular repercussions.

Involvement of the renal arteries is very characteristic of this syndrome and can be unilateral or bilateral. Therefore, the main symptom is renovascular hypertension Fig.2. Other disorders such as intestinal angina, intermittent claudication and heart failure are rare. It is important to rule out important organ damage so renal function tests, echocardiogram and fundus examination are always indicated.

Since most patients are children, an ultrasound - first approach is recommended Fig.3. Doppler ultrasound is of great help to detect critical stenosis in the renal flow. Spectral Doppler shows a Parvus tardus pattern in intrarenal arteries (small amplitude, prolonged systolic rise and low resistance index) Fig.4. This suggests a proximal stenosis of the renal artery. Unilateral or bilateral Parvus tardus waveform in kidneys in a child with hypertension should make suspect the possibility of this syndrome.

CT angiography (CTA) or magnetic resonance angiography (MRA) and arteriography are the imaging protocol of choice since they allow the visualization of the aorta and its main branches. They are also necessary for surgical planning. Multiplanar and three-dimensional reconstructions should be performed to know if there is involvement of renal arteries, superior mesenteric artery and celiac trunk and to define the extension and location of the stenosis Fig.5. Diameters of the aorta in the stenosed segments can be evaluated through these images Fig.6.

Diagnosis is usually confirmed with computed tomographic angiography (CTA) or magnetic resonance angiography (MRA). These techniques demonstrate narrowing of the abdominal aorta. Segmental aortic stenosis will be located at the inter-renal aorta in most of cases (19-52%) Fig.7. It is very frequent ostial stenosis of the renal arteries (60-90%), with less common involvement of the celiac trunk and the superior mesenteric artery (20-40%) Fig.8 and 9. Inferior mesenteric artery is rarely affected.
Conventional angiography is considered by some authors as the imaging technique of choice. However, techniques using MRI and CT have become equally valuable tools. Their large fields of view, as well as the fact that they are noninvasive, make them far more convenient in the pediatric age.

Although uncommon, MAS can manifest with stenosis of the thoracic aorta, even without involvement of the abdominal arteries Fig.10. Cases with stenosis of the lower thoracic and upper abdominal aorta have been described, with a variable length between 4 and 15 cm.

Fibrodysplasia of the intima and distortion of the internal elastic lamina without inflammatory changes are frequent histological findings. In the absence of signs of vasculitis, stenosis of the abdominal aorta at the level of the renal arteries suggests the diagnosis of MAS.

The treatment is medical and surgical. Medical measures are directed to control the symptoms that derive from hypertension and ischemic complications Fig.11. Sometimes they are not enough to control the disease and a surgical or endovascular approach is necessary. These will be indicated in case of uncontrolled hypertension and progressive renal failure despite an aggressive medical management Fig.12.

Surgical treatment should be individualized based on the severity of the symptoms and the age and size of the child. Several studies have found the aortic bypass as the procedure of choice due to its high success rate and resolution of the symptoms. Endovascular treatment through angioplasty and stenting have shown favorable short-term results, although evolution and long-term results still need to be assessed. Nevertheless, surgery requires a careful approach in the pediatric age and should, as far as possible, be delayed until the end of puberty.
Fig. 1: MR angiography. Mid-Aortic syndrome. Stenosis of the infrarenal abdominal aorta with concomitant stenosis of right renal artery.

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**Fig. 2:** Mid-Aortic syndrome in a 15-year-old patient with renovascular hypertension. Stenosis of the infrarenal abdominal aorta. Note that there is marked stenosis of the left renal artery. Stenosis at the ostium of the superior mesenteric artery (SMA) is also visualized.
Fig. 3: Decreased caliber of the infrarenal abdominal aorta. Note that visceral arteries (celiac trunk and superior mesenteric artery) and infrarenal abdominal aorta have the same diameter, a clearly pathological finding.

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Fig. 4: Mid-Aortic syndrome in a 4-year-old child. Parvus tardus waveform in intrarenal arteries, suggesting renal artery stenosis.

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Fig. 5: CT Angiography with three-dimensional reconstruction. Ostial stenosis of superior mesenteric and left renal artery. Stenosis in MAS typically involves the ostium of the vessel.

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Fig. 6: CT Angiography with multiplanar reconstruction. Abdominal aortic stenosis with minimum caliber at the level of renal arteries (Frequent finding in this syndrome).

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Fig. 7: Stenosis of the abdominal aorta at the level of the renal arteries. Most common location of the stenosis in the Mid-Aortic syndrome (MAS).

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Fig. 8: Ostial stenosis of a renal artery (aortic branch more frequently affected). No involvement of the celiac trunk and the superior mesenteric artery.

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Fig. 9: Idiopathic Mid-Aortic syndrome. There is an abrupt caliber change of the abdominal aorta below the level of the SMA origin. In this case, there is marked stenosis at the ostium of the SMA.

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Fig. 10: Thoracic aortic stenosis in a 11-year-old girl with Mid-Aortic Syndrome. The descending thoracic aorta, distal to the left subclavian artery, presents an irregular morphology and reduced caliber. Ascending thoracic aorta is normal.

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**Fig. 11:** Mid-Aortic Syndrome in a 2-year-old child with Mucolipidosis II. Renal artery stenosis with renal infarction.

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**Fig. 12:** Idiopathic Mid-Aortic Syndrome in a 13-year-old boy. There was severe stenosis of the right renal artery and renovascular hypertension difficult to control with medical therapy. The patient was taken up for hepatorenal bypass.

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Conclusion

MAS is an important cause of hypertension in childhood and a rare but potentially lethal entity. As most patients are children and young adults, the clinical benefits of early diagnosis and treatment are undebatable. Radiologists should understand the key radiologic findings and suspect this disease when it is present.


