

## Insights into cardiovascular involvement in patients with Cogan's syndrome

We read with interest the recent paper of Dr. Weyn and co-authors [1] on the treatment of a patient affected by Cogan's syndrome. The authors reported a young female patient with a history of interstitial keratitis and audiovestibular disease, presenting with supra-aortic vessel stenosis and with left main coronary artery occlusion.

We congratulate the authors on their work, and would like to add a few points:

1. The typical Cogan's syndrome, first described in 1945, is represented by an association of interstitial keratitis (IK) and vestibuloauditory symptoms [2]. In 1980, Haynes et al. [3] suggested that the definition of this condition should be enlarged to include patients with ocular symptoms other than IK or audiovestibular symptoms different from Meniere-like episodes, and proposed diagnostic criteria for typical and atypical Cogan's syndrome. This means that a large spectrum of clinical manifestations may be included in the definition of this syndrome. Furthermore, laboratory findings of inflammation or positivity for an autoimmunity disorder are merely indicative of disease activity, and at the moment there is no specific diagnostic or pathognomonic technique for Cogan's syndrome. We could say the same for the etiology of the disease.

The association with other systemic involvement and with the cardiovascular system in particular would raise the question of an overlap of Cogan's syndrome with other diseases. We mean in particular Takayasu's arteritis, affecting predominantly the large arteries, with narrowing, occlusion or dilatation of the involved segments. A 'triphasic' pattern of disease progression has been described [4]. Phase 3 of Takayasu's arteritis is the late fibrotic stage, in which stenoses of the aorta and its major branches predominate. Takayasu's arteritis should not be considered as confined only to young Oriental women, since the disease is becoming increasingly recognized worldwide. A North American study [5] suggested an incidence similar to that in Japan. The overlap in classification criteria may be easily seen in vasculitis. In particular, the overlap between

Takayasu's and the inflammatory ocular and auditory features of Cogan's syndrome has already been described in the literature [6] and should be considered as well in the case reported by the authors.

2. We agree with the authors on the surgical management of this patient. Unfortunately, such patients, as usually happens with Takayasu's disease, even if young, often do not benefit from left internal thoracic artery (LITA) revascularization due to lesions on the subclavian artery, and due to the hypothetical inflammatory involvement of the LITA itself.

However, using venous bypass, it should be remembered that the proximal anastomosis is performed on a diseased aorta with some grade of inflammation, often with large thickness, and this could predispose to intimal proliferation and graft occlusion. A discrepancy may also exist between aortic thickness and the anastomosed saphenous vein. For this we believe that in such patients veins of larger calibre should be used, taking the veins from the upper thigh and not from the leg.

3. The third issue we would raise is about the choice of valve prosthesis in these patients. Some time ago, we treated a 34 year old Caucasian woman with a previous diagnosis of Cogan's syndrome. The patient developed deafness and a reduced visual field despite chronic treatment with steroids and azathioprine. She was positive for ANA antibodies. She was referred to our observation for aortic valve stenosis with a mean gradient of 80 mm Hg. The choice of valve prosthesis in this clinical scenario may be controversial. In fact, we would usually prefer to implant a mechanical valve in a young patient. On the other hand, we must take into account that these patients are usually on long term treatment with steroids and other immunosuppressive agents, which could interfere with anticoagulation.

In this particular case we implanted a biological valve, in line with a previous report from Paolini et al. [7].

We are aware of the current controversy concerning the treatment of this syndrome. The lack

of standardized protocols at this time is due mainly to the rarity of this pathology.

We congratulate again Dr. Weyn and co-authors on their work.

## References

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## Author's response

We would like to thank Dr. Lentini and co-authors for their interest and remarks on our case report “Cogan's syndrome with left main coronary artery occlusion”. We would like to respond to their letter.

1. The diagnosis of Cogan's syndrome (CS) is based upon the presence of inflammatory eye disease and audiovestibular dysfunction. In 1980 Haynes et al. [1] proposed retaining the diagnosis of atypical CS when different ocular and audiovestibular symptoms were observed. The modifying terms “typical” and “atypical” CS have been abandoned because this distinction no longer appears to carry prognostic significance. We agree with the authors that the large-vessel vasculitis associated with CS may also resemble Takayasu's arteritis. Our case report indeed illustrates the overlap between Takayasu's arteritis and the inflammatory ocular and auditory features of CS.
2. Coronary revascularisation surgery in Takayasu's arteritis and CS with large-vessel vasculitis is often complicated due to stenosis or

occlusion of the innominate and subclavian arteries, as they reduce the availability of the internal thoracic arteries as coronary bypass grafts [2]. Moreover, active inflammation of the aorta may require the use of an artificial patch at the side of the anastomoses of the vein grafts with the aorta. Revascularisation procedures are often unsuccessful when performed in patients whose inflammation is uncontrolled. Perioperative administration of steroids may be important to decrease inflammation.

3. Aortic insufficiency due to aortitis is the most commonly occurring cardiac valvulopathy in CS (15%); mitral insufficiency is reported more rarely. The mechanism of aortic insufficiency is generally thought to be a combination of dilatation of the aortic root at the aortic valve and thickening of the valve cusps. Aortic valve replacement with or without aortic root replacement is the only curative treatment, but frequently gives rise to cardiovascular complications due to the extensive and severe lesions and also due to the necessity to manipulate

fragile and inflamed tissue. Complications include detachment of the prosthetic valve, anastomotic aneurysm, infective endocarditis, hemorrhage and cerebrovascular disease [3]. When valve replacement is performed, either a mechanical or a bioprosthetic valve can be used. Based on our experience, we would choose a mechanical valve in patients with a projected long life-span because of greater durability and improved patient survival at 15 years, despite the use of steroids and immunosuppressive agents which could interfere with oral anticoagulation [4].

## References

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