The other symptoms and signs are not always present and their absence does not rule out ACS. Edema is an early symptom and the paresthesia and paralysis are delayed signs (caused by nerve ischemia). In ACSh, the hand is characteristically in flexion, due to the intense pain caused by movement/extension of the fingers. The definitive treatment is fasciotomy, which is almost always performed. When the clinical findings are insufficient or to decide when to perform a fasciotomy, direct measurement of the intracompartmental pressure is an option.[1]

The development of ACS after transradial catheterization has been described in the forearm. This part of the upper limb contains 3 compartments (anterior, posterior, and mobile wad); ACS affects the anterior compartment (also known as the volar compartment) because the radial artery passes through this compartment. This particular ACS has an incidence of about 0.125%. It is generally caused by perforation (by guidewires or catheters) of the small arteries of the forearm proximal to the PS, and the bleeding occurs at a distant site from the PS. In contrast to the "classic" ACS, if it is identified at an early stage, ACS of the forearm can usually be resolved using an elastic tensor bandage or by inflating a pressure cuff on the affected forearm.[2] In refractory cases, a fasciotomy should be performed. Early treatment failure can lead to a chronic deformity called Volkmann ischemic contracture.

An ACSh after a transradial procedure is highly unusual. In our institute, this is the first case among 17 965 transradial procedures, giving an incidence of 0.005%. The hand has 11 compartments (thenar, hypothenar, central palmar, adductor, and 7 interosseous spaces). The "classic" treatment recommends removal of the compression bandages and a fasciotomy. In the only other previously described case of ACSh, spontaneous laceration of the hand occurred and the ACS was resolved with drainage and surgical debridement.[3] Our exceptional case of ACSh after transradial catheterization was easily resolved by expanding the PS hole using a scalpel, with no need for surgery. We believed that the ACSh was caused not by artery rupture, but by inadequate initial compression and the subsequent hematoma, which further complicated the compression. We also considered that, due to the small initial transradial PS, the blood was unable to drain to the exterior and preferentially drained into the hand. Accordingly, use of a scalpel to simply expand the puncture hole in the epidermis and the subcutaneous cellular tissue would facilitate blood outflow and complete resolution of the ACS. Due to its simplicity, when ACSh develops after transradial catheterization, this maneuver should first be attempted before fasciotomy is performed.

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Prevalence of Neuroendocrine Tumors in Patients With Cyanotic Congenital Heart Disease

Prevalencia de tumores neuroendocrinos en pacientes con cardiopatías congénitas cianóticas

To the Editor,

Pheochromocytoma and paraganglioma are neuroendocrine tumors (NETs) that produce catecholamines derived from neural crest cells, localized in the adrenal medulla (90%) or in extra-adrenal chromaffin tissue (10%). Their prevalence is between 0.2% and 0.6% in hypertensive adults, 5% in adrenal incidentalomas, and 0.05% to 0.1% in autopsy series.[1] Although they usually present as isolated tumors, they can be associated with hereditary syndromes such as multiple endocrine neoplasms, neurofibromatosis, or von Hippel Lindau syndrome. Furthermore, there have been case reports and small series highlighting the association between cyanotic congenital heart defects (CCHD) and NETs.[2] Recently, Opotowsky et al.[3] reported an increased risk of NETs in patients with CCHD in a multicenter study in which the role of chronic hypoxia in association with genetic susceptibility was proposed as the underlying pathogenic mechanism for these tumors.

A retrospective analysis was performed of 3311 adults with congenital heart defects, 173 with CCHD, and 33 with Eisenmenger syndrome in a national referral center for adult congenital heart defects. The median length of follow-up was 25 years (range, 10.5 years). All patients with suspected NET under follow-up in the endocrinology department underwent computed tomography and metaiodobenzylguanidine scintigraphy, with monitoring of catecholamines in urine.

A total of 8 NETs were identified in 7 patients with CCHD (4.6%) (Table 1). Overall, 48.8% were men and the median age was 40.0 years (range, 19.0-47.0 years). All patients had active cyanosis at the time of diagnosis (mean, 36.0 ± 11.3 years), including 1 patient with Fontan circulation with venovenous collaterals. The mean baseline arterial oxygen saturation was 83.4% ± 6.3%, mean hemoglobin was 18.1 ± 2.0 g/dL, and mean hematocrit was 66.5% ± 7.3%. Three patients had Eisenmenger syndrome (9.1% of all patients had this syndrome). On analysis of all patients with congenital heart defects, including those with noncyanotic disease, 1 additional NET was identified in a patient with partial anomalous pulmonary venous drainage (0.2% overall).

Table 1
Characteristics of Patients With Congenital Heart Defects and Neuroendocrine Tumors

Congenital heart defect	Prior surgery	Age, y	Sex	Baseline saturation, %	Eisenmenger syndrome	Neuroendocrine tumor	Catecholamines in urine	Tumor resection
Ebstein Anomaly, SVC drainage to LA	No	40	F	92	No	Pheochromocytoma	Yes	Open-heart surgery
Double inlet single ventricle	No	42	F	82	Yes	Retroperitoneal paraganglioma	Yes	Open-heart surgery
Functional single ventricle by VSD	No	47	M	80	Yes	Glomus caroticum	No	Surgery
Pulmonary atresia with intact septum	Total cavopulmonary shunt	19	M	92	No	Pheochromocytoma	Yes	Open-heart surgery
TGA with VSD and pulmonary atresia	Glenn-type cavopulmonary shunt	39	M	75	No	Glomus caroticum	No	Pending
Double inlet single ventricle, D-TGA	Pulmonary banding, Glenn type cavopulmonary shunt	30	F	82	No	Retroperitoneal paraganglioma	Not available	Open-heart surgery
Single arterial outflow, ASD, VSD, situs inversus	Waterston fistula	41	F	81	Yes	Pheochromocytoma	Yes	Laparoscopy
						Carotid glomus	No	No

ASD, atrial septal defect; F, female; LA, left atrium; M, male; SVC, superior vena cava; TGA, transposition of the great arteries; VSD, ventricular septal defect.

Among patients with CCHD, 3 cases of pheochromocytoma (2 in the left adrenal gland and 1 in the right adrenal gland) and 5 cases of paraganglioma were diagnosed (3 cases of glomus caroticum [GC] and 2 cases of extra-adrenal paraganglioma in the retroperitoneal space). One patient (14.3%) had a pheochromocytoma and as well as GC. No other multisystemic syndromes associated with NET were detected. The form of presentation was hypertension in 3 patients with pheochromocytoma, abdominal pain in 1 patient with retroperitoneal paraganglioma, and hypoacusia in 1 patient with GC; diagnosis was coincidental in 2 patients. Catecholamines were detected in urine in all patients with pheochromocytoma and in the 1 patient with retroperitoneal paraganglioma, with elevated metanephrine in 3 patients (50%), elevated normetanephrine in 3 patients (50%), and elevated vanilmandelic acid in 2 patients (33.3%) (Table 2). Three patients (60%) had simultaneous elevation of at least 2 metabolites in urine. Data were not available for 1 patient with retroperitoneal paraganglioma because the diagnosis was made at a different hospital.

After confirmation of the diagnosis, the tumor was resected by urologists, ear-nose-throat specialists, or general surgeons after treatment with alpha blockers and infusion of saline solution. In the patient with 2 tumors, only the pheochromocytoma was resected as a conservative approach was chosen with the GC. Postoperative complications included acute pulmonary edema in a patient with pheochromocytoma who had fluid overload and transient ischemic attack, without sequelae, in another patient with GC. No patients died in hospital or during follow-up. In the histopathological analysis, only 1 sample showed findings suggestive of malignancy. None of the patients showed metastasis.

Here, we report our experience with NET in patients with CCHD in the largest Spanish series published to our knowledge. An increased prevalence of NET was observed, especially in patients with Eisenmenger syndrome, probably due to chronic exposure to hypoxia, which stimulates formation of erythropoietin and growth factors regulated by hypoxia inducible factor 1 (vascular endothelial growth factor and platelet-derived growth factor), thus favoring tumor genesis.[4] Our patients included those with uncorrected congenital heart defects, palliative corrections, and venovenous collateral flow after a Fontan procedure. All had active cyanosis at the time of diagnosis, defined as clinical cyanosis, and all had low baseline arterial oxygen saturation (in most patients $< 83\%$). In addition to hypoxia, genetic susceptibility has also been reported as a triggering factor, although in our series, only 1 patient had multiple NETs and the incidence (14.3%) was significantly lower than that published in previous series (up to 39%).[3]

Table 2
Detection of Catecholamines in Urine

Congenital heart defect	Tumor	Metanephrine	Normetanephrine	Vanilmandelic acid
Ebstein Anomaly, SVC drainage to LA	Pheochromocytoma	+	+	–
Double inlet single ventricle	Retroperitoneal paraganglioma	–	+	–
Functional single ventricle by VSD	Glomus caroticum	–	–	–
Pulmonary atresia with intact septum	Pheochromocytoma	+	–	+
TGA with VSD and pulmonary atresia	Glomus caroticum	–	–	–
Double inlet single ventricle, D-TGA	Retroperitoneal paraganglioma	Not available	Not available	Not available
Single arterial outflow, ASD, VSD, situs inversus	Pheochromocytoma, glomus caroticum	+	+	+

ASD, atrial septal defect; LA, left atrium; SVC, superior vena cava; TGA, transposition of the great arteries; VSD, ventricular septal defect.

The onset of NET can lead to general and hemodynamic deterioration in these patients and therefore early diagnosis and treatment are essential to reduce the risk of complications. The signs and symptoms of these tumors overlap with those associated with complications of CCHD, such as arrhythmias, hypertension, and heart failure. Thus, the presence of NET should be suspected with the onset of new symptoms in patients with CCHD, even after surgical correction, as these tumors are a potentially treatable cause of clinical deterioration in these patients. A multidisciplinary approach, with tumor resection in a specialized center, is associated with a high success rate, even in this population at risk.[2] This treatment is effective and is associated with good short- and long-term prognosis.

CONFLICTS OF INTEREST

Á. Sánchez-Recalde is an Associate Editor of *Revista Española de Cardiología*.

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Stented Bovine Jugular Vein Graft (Melody Valve) in Mitral Position. Could Be an Alternative for Mechanical Valve Replacement in the Pediatric Population?

Prótesis de yugular bovina con stent *(Melody) en posición mitral. ¿Posible alternativa a la prótesis mecánica en población pediátrica?*

To the Editor,

Congenital mitral valve disease is an uncommon condition. Medical treatment can be very complicated in some cases, leaving surgery as the only option. Surgical valvuloplasty often fails in children, especially in neonates and young infants, due to the presence of dysplastic valves with a small annulus and special anatomic features. In such cases, valve replacement is generally the only solution. We present 3 cases of Melody valve implantation in the mitral position.

Patient 1 was a 4-month-old infant weighing 4.6 kg with severe mitral regurgitation (MR) (valve with thickened leaflets, reduced mobility, and absence of central coaptation; annulus of 15 mm) that was refractory to medical treatment. Following Kay-Wooler annuloplasty, the boy showed moderate residual MR and was extubated, but he developed severe MR 14 days later and required ventilatory support. We decided to implant a Melody valve in the mitral position using the Boston technique[1] with some modifications.[2] Before initiation of extracorporeal circulation, the valve was expanded to 18 mm and a 3-mm pericardial sewing cuff was added to the center of the stent using loose sutures anchored to the strut chordae; the triangular struts at the proximal and distal ends of the stent were bent outwards, but the 3 struts supporting the valve commissures were left intact. The mitral valve was exposed using a superior transseptal approach. The posterior leaflet and its subvalvular apparatus and part of the anterior leaflet were resected, sparing the anterosuperior zone with its attachments to the anterior papillary muscle. The mitral prosthesis was crimped (6 mm) and attached to the posterior wall of the left ventricle to prevent left ventricular outflow tract (LVOT) obstruction during systole. The pericardial cuff was sutured to the native annulus and the valve was inflated to 4 atm with an 18-mm balloon (annulus diameter + 1). The cuff was tied down and the interatrial septum was reconstructed with a fenestrated pericardial patch (Figure 1). An intraoperative transesophageal echocardiogram (TEE) showed grade III periprosthetic MR. The valve was reinflated with a 22-mm balloon, and the outcome was favorable (grade I-II MR). No postoperative complications were observed and the patient was asymptomatic 9 months later. The echocardiogram revealed a mean mitral valve gradient of 3.6 mmHg and grade II periprosthetic MR. No LVOT obstruction was noted.

Patient 2 was a 7-month-old girl weighing 4.7 kg, who had been treated for complete atrioventricular canal defect at another hospital using the double-patch technique with cleft closure and the Alfieri technique, following pulmonary artery banding. Postoperative clinical course was indolent and the patient required prolonged hospital stay. The infant had a severe double mitral valve lesion and an annulus of 15 mm. She failed to thrive and developed heart failure despite maximum medical treatment. It was decided to implant the Melody valve in the mitral position using the technique described above, with expansion of the prosthesis to 17 mm. Intraoperative TEE showed no evidence of residual MR or LVOT obstruction (Figure 2). The patient was asymptomatic at the 7-month follow-up visit and had no residual lesions (mean gradient, 3 mm Hg; no MR).

Patient 3 was a 3-kg neonate with congenital aortic valve stenosis (peak gradient, 100 mm Hg). Valvuloplasty had been performed when the child was 2 days old, but the next day, extracorporeal membrane oxygenation was required due to ventricular dysfunction. The neonate was weaned off the oxygenation system after 5 days. The echocardiogram showed a residual aortic stenosis of 50 mmHg, a patent foramen ovale of 4 to 6 mm with a significant left-to-right shunt and moderate MR with