

Aorto–Ventricular Tunnel

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Abstract

Aorto-ventricular tunnel is a congenital, extracardiac channel which connects the ascending aorta above the sinutubular junction to the cavity of the left, or (less commonly) right ventricle. There may be associated defects of the proximal coronary arteries, or the aortic or pulmonary valves. Occasional patients present with an asymptomatic heart murmur and cardiac enlargement, but most suffer heart failure in the first year of life. The etiology of aorto-ventricular tunnel is uncertain. It appears to result from a combination of maldevelopment of the cushions which give rise to the pulmonary and aortic roots, and abnormal separation of these structures. Its incidence is also uncertain, estimates ranging from 0.46% of fetal cardiac malformation to less than 0.1% of congenitally malformed hearts in clinico-pathological series. Optimal management of aorto-ventricular tunnel consists of diagnosis by echocardiography, supplemented with cardiac catheterization as needed to elucidate coronary arterial origins or associated defects, and prompt surgical repair.

Keywords

Aorto-ventricular tunnel, aortico-left ventricular tunnel, aortico-right ventricular tunnel, congenital heart malformation, neonatal heart failure, aortic stenosis, congenital coronary artery stenosis, coronary artery atresia, aortic insufficiency, aortic regurgitation, pulmonary stenosis

Disease name / Synonyms

In their original description of the malformation, Edwards and Burchell (Edwards and Burchell, 1957) considered the malformation a “separation between the aorta and the heart”, or type of aneurysm which “lay against the outflow tract of the right ventricle and origin of the pulmonary trunk”. The term “aortico-left ventricular tunnel” was used subsequent to Levy’s publication in 1963 (Levy *et al.*, 1963), and “aorto-left ventricular tunnel” was later introduced by Ross and colleagues (Somerville *et al.*, 1974). Recognizing that the tunnel may extend to either the left or the right ventricular cavity, the more

general name “aorto-ventricular tunnels” has recently been applied to this group of malformations (McKay *et al.*, 2002). The defect is not a component of any described genetic syndrome.

Definition / diagnostic criteria

An aorto-ventricular tunnel is an extracardiac channel which connects the ascending aorta above the sinutubular junction to the cavity of the left or right ventricle. Among 130 cases reported in the literature, more than 90% communicated with the left ventricle. It differs from a ruptured sinus of Valsalva aneurysm (sinus of Valsalva fistula) in having its vascular orifice in the

tubular aorta, rather than a sinus of the aortic valve, and in passing outside the heart into the tissue plane between the muscular subpulmonary infundibulum and the aortic sinuses. The aortic opening of most tunnels lies above the right coronary sinus of Valsalva. In these cases, the tunnel virtually always communicates with the left ventricle in the fibrous triangle beneath the left - right coronary commissure, or the right ventricle immediately above or below the subpulmonary infundibulum. In aorto-left ventricular tunnel, the right coronary aortic leaflet is thus unsupported for a variable portion of its hinge-point and may appear to arise from a bar of fibrous tissue spanning the aortic root (Cook *et al.*, 1995). Tunnels lying above the left sinus of Valsalva or the intercoronary commissure have less uniform morphology and enter the left ventricle further away from the aortic valve, apparently through infoldings of fibrous tissue. It is extremely rarely, if ever, that an aorto-ventricular tunnel passes through intracardiac myocardium to reach the cavity of the ventricle, a feature which serves to differentiate it from coronary-cameral fistula (McKay *et al.*, 2002).

The ostium of a coronary artery may lie within an aorto-ventricular tunnel, and absence of the origin of the left (Saylam *et al.*, 1974) or right (Bove and Schwartz, 1967; Horváth *et al.*, 1991; Hovaguimian *et al.*, 1988; Rauzier *et al.*, 1997; Rosengart *et al.*, 1993; Sommerville *et al.*, 1974) has been observed with this anomaly. Associated lesions of the aortic valve occur in about 20% of patients, ranging from two-leaflet valves without obstruction (Edwards and Burchell, 1957; Levy *et al.*, 1963) to severe dysplasia or atresia (Bitar *et al.*, 1993; Guyton *et al.*, 1986; Sousa-Uva *et al.*, 1996). In addition, older patients may acquire leaflet perforation (Meldrum-Hanna *et al.*, 1986; Warnke *et al.*, 1988) or aortic incompetence (Akalin *et al.*, 1989) as the result of hydrodynamic trauma to the unsupported leaflet or aortic dilatation. Stenosis of the pulmonary valve (Jureidini *et al.*, 1989; Martin-Jimenez *et al.*, 1996) occurs less frequently (around 5% of reported cases), while compression of the right ventricular outflow tract by the tunnel may produce subpulmonary obstruction (Knott-Craig *et al.*, 1992). Rarely, both valves are stenotic (Levy *et al.*, 1963; Turley *et al.*, 1982).

Histologically, the arterial end of the tunnel resembles the aorta with fibrous tissue, elastic fibers and smooth muscle cells, while the ventricular end contains hyalinized collagen and muscle, reflecting that the "walls" of tunnels incorporate the structures through which they pass. Within the tunnel itself, there may be a well-defined junction between ventricular and

arterial components, in addition to cystic or membranous structures reminiscent of cardiac valve leaflets (Kleikamp *et al.*, 1992).

Differential diagnosis

Aorto-ventricular tunnel is to be distinguished from other lesions which cause rapid run-off from the aorta and produce cardiac failure. These include sinus of Valsalva fistula, common arterial trunk with valvar regurgitation, aorto-pulmonary window, ventricular septal defect with aortic regurgitation, persistent patency of the arterial duct, coronary-cameral fistula, valvar aortic stenosis and regurgitation, and cerebral arterio-venous malformation. Because of its "to-and-fro murmur", tetralogy of Fallot with absent pulmonary valve can also mimic an aorto-ventricular tunnel in which there is associated right ventricular outflow obstruction.

Etiology

While the etiology of aorto-ventricular tunnel is unknown, the substrate for its formation and that of the associated valvar and coronary arterial lesions may be inferred from developmental anatomy (Bernanke *et al.*, 2002; Bogers *et al.*, 1998; Ya *et al.*, 1998). The cushions which form the facing aortic and pulmonary sinuses with their respective valvar leaflets normally become separated by an extracardiac tissue plane, due to regression of surrounding muscle. The coronary arteries, also initially encased by this cuff of myocardium, grow through it to connect with the aortic sinuses. Failure of this tissue plane to develop normally might then result in a tunnel above one of the facing aortic sinuses and explain also the potential involvement of the proximal coronary arteries and valve leaflets. This produces one of the few congenital malformations which may involve both the pulmonary and aortic valves.

Clinical description

A loud "to-and-fro" murmur, usually with systolic and diastolic thrills, invariably radiates over the entire precordium in aorto-ventricular tunnel, and bounding pulses indicate rapid aortic run-off. In older patients, these signs may suggest aortic valve stenosis with incompetence, but the second heart sound should have a normal aortic component. Symptoms of heart failure can present at any time, although most patients become symptomatic during the first year of life. The severity and progression of heart failure is also variable, ranging from many years of asymptomatic compensation (Akalin *et al.*, 1989; Kafka *et al.*, 1989; Ribeiro *et al.*, 1985; Serino *et al.*, 1983) to rapid decompensation (Bove and Schwartz, 1976; Palacio *et al.*, 1964), sudden death (Roberts and Morrow, 1965), or death *in utero* (Sousa-Uva *et al.*, 1996). This may reflect variable compression of coronary arteries or

obstruction to the right ventricular outflow tract, although it has not, in general, been possible to correlate clinical course with specific morphology of the tunnel. The exceptions are aorto-right ventricular tunnel with pulmonary stenosis, in which the onset of heart failure is delayed (Hruda *et al.*, 2002), and tunnels with severe associated aortic valve obstruction. In this latter group, congestive heart failure, with or without low cardiac output, supervenes early, and nearly one third of reported cases have died before birth or on the first day of life.

Diagnostic methods

Echocardiography is the diagnostic investigation of choice (Bash *et al.*, 1985; Cook *et al.*, 1995; Grant *et al.*, 1985; Humes *et al.*, 1986; Perry *et al.*, 1983; Sousa-Uva *et al.*, 1996; Sreeram *et al.*, 1991). Transthoracic cross-sectional imaging in a parasternal long-axis view demonstrates the tunnel itself, as well as its aortic origin and left ventricular opening. On color-Doppler studies, diastolic flow is seen passing from the aorta to the left ventricle, and systolic, from the ventricle to the aorta. Tunnels which open into the right ventricle are visualized in the short axis view, while left ventricular function, which may be variably impaired with hypertrophy and dilatation, is assessed in short axis cuts. Magnetic resonance angiography also has been used to demonstrate tunnels to the left (Humes *et al.*, 1986) and right (Hruda *et al.*, 2002) ventricles but is not readily available in clinical practice. Cardiac catheterization with angiography is now indicated only when associated lesions or coronary arterial origins cannot be evaluated with certainty on noninvasive studies.

Epidemiology

The incidence of aorto-left ventricular tunnel has been estimated to be around 0.1% of congenitally malformed hearts from review of clinical and pathological material (Okaoroma *et al.*, 1976), and 0.46% of cardiac malformations identified by fetal echocardiography (Cook *et al.*, 1995). About twice as many cases have been reported in males as in females, but it is seldom seen in patients of Asian, Oriental, or African descent. Although extremely rare, aorto-ventricular tunnel is the most common cause of abnormal blood flow from the aorta to a ventricle in infancy.

Antenatal diagnosis

It is possible to reliably diagnose aorto-ventricular tunnel on fetal echocardiography after 18 weeks gestation. Hypertrophy and dilatation of the left ventricle with progressive reduction of its shortening fraction are consistent features, and there is often disproportionate dilatation of the aortic root with apparent incompetence of the valve. Using color flow Doppler imaging, blood

flow around the aortic valve has been demonstrated (Sousa-Uva *et al.*, 1996). There are no specific molecular markers for aorto-ventricular tunnel, and it is not associated with any recognized genetic syndrome.

Management

Surgical correction should be undertaken without delay, even in asymptomatic patients, as only those repaired in the first six months of life have been shown to have subsequent normalization of left ventricular size and function. Repair consists of closing the tunnel such that the aortic valve is supported, the coronary circulation is not compromised, and left or right ventricular outflow obstruction is prevented or relieved. In most cases, this has been accomplished by transaortic patch closure of the aortic end, and placement of a second patch through the tunnel itself to close the ventricular orifice and support the aortic valve. Alternatively, the tunnel wall can be used to achieve an equivalent anatomical result (Grünenfelder *et al.*, 1998). Simple suture closure of the aortic orifice has occasionally given good results (Norwicki *et al.*, 1977; Spooner *et al.*, 1978), but more often, the tunnel recurs or progressive regurgitation through an unsupported or distorted leaflet leads to subsequent aortic valve replacement. If the ventricular end of an aorto-left ventricular tunnel is not closed, residual high pressure in the blind-ending pouch may compress the right ventricular outflow (Knott-Craig *et al.*, 1992).

When the ostium of a coronary artery arises proximally within a tunnel, the patch is deviated distally to conserve perfusion from the aorta. More distal origin of a coronary artery from a tunnel above the right aortic sinus is managed by resection of the orifice and reattachment to the ascending aorta (Grünenfelder *et al.*, 1998; Hucin *et al.*, 1989; Rauzier *et al.*, 1997). Distal coronary origin in a tunnel arising above the left aortic sinus is more difficult to manage, because it lies behind the heart. As these generally have been associated with tunnels to the right ventricle, however, closure of just the ventricular end is an option to maintain coronary perfusion (Hruda *et al.*, 2002).

Associated lesions of the aortic valve are treated as indicated at the time of tunnel repair. This has included commissurotomy (Diamant *et al.*, 1985; Villani *et al.*, 1980; Webber *et al.*, 1991), homograft root replacement (Weldner *et al.*, 1996), or aortoventriculoplasty (Guyton *et al.*, 1986) for stenosis or atresia in neonates or small infants, as well as repair or replacement of the valve in older patients. Obstruction of the pulmonary valve has been successfully managed by percutaneous valvoplasty preoperatively (Martin-Jimenez *et al.*, 1996) or open valvotomy at the time of surgery (Hruda *et*

al., 2002; Turley *et al.*, 1982). However, attempted percutaneous balloon dilation did not relieve the obstruction on one occasion (Hruda *et al.*, 2002).

Transcatheter closure of a tunnel to the left ventricle with an Amplatzer duct occluder has been reported in a single patient (Chessa *et al.*, 2000), but attempted coil closure of one to the right ventricle was not effective (Hruda *et al.*, 2001). Given the desirability of supporting the aortic leaflet and the variable origins of coronary arteries in this malformation, it is questionable if percutaneous interventions can achieve long-term outcomes equivalent to those of current surgical techniques, for which operative mortality approaches zero (Horváth *et al.*, 1991).

Unresolved questions

The very long-term results of two-patch repair in the modern era are awaited, as are elucidation of the molecular or genetic basis of the malformation.

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