as previously defined. Repeat blood cultures during therapy are important to determine the effectiveness of treatment.

REFERENCES


Double Outlet Right Ventricle with Absent Aortic Valve*

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A case of double outlet right ventricle with multiple associated cardiovascular anomalies, including total absence of the aortic valve, is reported.

Double outlet right ventricle (DORV) is the pathologic result of aberrant trunco-conal cardiovascular development. The basic anatomy of this unusual form of congenital heart disease, as well as the associated cardiovascular anomalies, have been well described.1-3. The purpose of this communication is to describe an associated absence of the aortic valve, which has not, to our knowledge, been previously reported.

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Case Report

A 7 lb 6 oz term caucasian boy was transferred to Denver Children's Hospital at 36 hours of life because of severe cyanosis and cardiomegaly. On admission, he was tachypneic and the peripheral pulses were diminished. Pertinent cardiac findings included right ventricular lift; split first heart sound; loud second heart sound; and a rough, low-pitched systolic murmur in the left axilla, associated with a softer, high-pitched diastolic murmur. At the lower left sternal border, the murmur became "to-and-fro."

A complete blood count, serum electrolytes, serum glucose, and blood urea nitrogen were normal. Arterial blood gas analysis on FIO2 80 percent revealed PO2 55 mm Hg, PCO2 27 mm Hg, and pH 7.41. The electrocardiogram showed a heart rate of 135 beats/min, mean frontal QRS axis +90°, P-R interval 0.14 sec, right and left atrial enlargement, and severe right ventricular hypertrophy. Chest x-ray examination demonstrated marked cardiomegaly and pulmonary vascular congestion. Echocardiogram was technically inadequate to support a specific anatomic diagnosis. However, it did demonstrate a dilated right ventricular (RV) and diminutive left ventricular (LV) cavity. The tricuspid valve (TV) had a wide excursion, but the mitral valve (MV) appeared to have markedly restricted motion. A great vessel was anteromedially located, but the relationship to the interventricular septum and atroventricular valve could not be demonstrated.

The infant subsequently underwent cardiac catheterization which demonstrated markedly elevated RV (90/0/10 mm Hg), right (RA) and left (LA) atrial pressures (a = 12, v = 8, m = 9 mm Hg in both). The catheter could not be made to enter either great vessel or the LV. Biplane cineangiograms were performed in the RV and LA which demonstrated complete passage of contrast medium from LA to RA and then to the RV. Filling of both great vessels from the RV occurred with aortic (Ao) opacification slightly delayed. A diagnosis of DORV with mitral atresia was made. During the procedure, the infant suffered respiratory arrest, requiring vigorous resuscitation.

Following the procedure, the infant's condition further deteriorated and death occurred despite all supportive measures. Post-mortem examination confirmed the diagnosis of DORV. Specifically, the heart was markedly enlarged—principally the right atrium and right ventricle. The aorta arose completely from the right ventricle. The former was dis-
placed anteriorly and to the right and was rotated so that the
coronary arteries arose from the left anterior and posterior
positions. Within the aorta, three tiny ridges were seen
beneath each coronary ostium and on the opposing wall. No
valve tissue was present, but the level of the ridges was
elevated to a position similar to that of the pulmonic valve
(Fig 1). Approximately 5 mm beneath the ridges, a small
mass of extremely dysplastic tissue arose from the anterior
base of the aorta and was attached to the septal leaflet of the
tricuspid valve. The pulmonic valve was normal and the
pulmonary artery larger than the aorta (15 mm vs 9 mm
diameter). Associated cardiovascular anomalies included a
fossa ovalis atrial septal defect (ASD), patent ductus ar-
teriosus (PDA), thickened and dysplastic but normally posi-
tioned TV, and a rightwardly displaced and hypoplastic
mitral valve with a double ostium. In addition, although a
blunt probe could be passed from the MV to the RV, entering
beneath the aorta, no adequate LV could be found. The only
noncardiac anomaly was an accessory spleen.

**DISCUSSION**

DORV can be divided into several types depending on the position of the ventricular septal defect (VSD),
the presence or absence of associated pulmonic stenosis
(PS), and the presence of associated major anomalies
such as total anomalous pulmonary venous return,
common atrioventricular valve orifice, or mitral stenosis
or mitral atresia. Other associated anomalies include
ASD (fossa ovalis type), persistent left superior vena
cava, PDA, absent or bicuspid pulmonic valve (PV),
coarctation of the aorta, MV abnormalities, hypoplasia of the
LV, partial anomalous pulmonary venous return, and asplenia. Our patient clearly had DORV, based on the position
and rotation of the aortic annulus.

The mitral valve was hypoplastic, with a double
ostium. The LV cavity was extremely small and the VSD
was found beneath the aorta. However, PS was not
present. Other abnormalities included the ASD and
PDA.

Of particular interest was the total absence of the
aortic valve. The ridges mentioned above presumably
represented the valve annulus. The dysplastic tissue
attached to the aorta clearly arose well beneath the
valve, and presumably acted to impede the egress of
blood through the aorta—as the latter was seen by cine-
angiography to opacify later than the pulmonary artery.
Free aortic insufficiency must have contributed heavily
to the infant’s demise.

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**Volume-controlled Lung Lavage in a
Woman with Cystic Fibrosis**

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A 22-year-old woman with cystic fibrosis was treated by
volume-controlled lavage of each of her lungs on
two occasions. Following the first lavages, the patient’s
vital capacity increased by 1.03 liters and her 1-sec
forced expiratory volume increased by 0.70 liters/sec-
ond. Similar improvements, although less pronounced,
were noted after both lungs had been lavaged a second
time. It is concluded that volume-controlled lung lavage
can be of benefit as an adjunct in the treatment of pa-
tients with cystic fibrosis.

**Cystic fibrosis** (mucoviscidosis) is a genetic disease of
the exocrine glands in which viscous mucus tends to
obstruct peripheral airways and predisposes to inflamma-
tion of lung tissues. Volume-controlled lung lavage has
been reported to be effective in removing impacted
secretions from the airways and to result in clinical
improvement. Braunstein and Fleegler, reported that volume-controlled lung lavage produced an acute
deterioration in the clinical and physiologic status of
a 24-year-old cystic fibrosis patient, from which he
had not recovered one month later. Reported here are
the results obtained with volume-controlled lung lavage
in a 22-year-old woman with cystic fibrosis.

**CASE REPORT**

The patient was first seen in October, 1972, at age 21. She
complained of a chronic, productive cough, blood-streaked
sputum, exertional dysnea, and denied past or present
gastrointestinal symptoms. Bilateral apical crepitations which
cleared with coughing were heard on auscultation of the
chest. The remainder of the physical examination was nor-
mal. Chest x-ray films revealed prominent bilateral posterior
apical densities. *Pseudomonas aeruginosa* was present in the
patient’s sputum. A diagnosis of chronic bronchitis was made
and the patient was treated with cephalothin, nebulized
racemic epinephrine, and postural drainage.

When the patient was seen in February, 1973, pulmonary
function tests revealed a decrease in vital capacity (VC) and
1-sec forced expiratory volume (FEV1), as well as an
increase in the residual volume (RV) (Table 1). Broncho-
grams revealed bilateral apical bronchiectasis. Four sweat
tests done by pilocarpine iontophoresis on 0.27-0.41 g of
sweat yielded values of 97-102 mEq/L of chloride (normal
upper limit, 60 mEq/L). The serum carotene level and
quantitative stool fat determinations were within normal
limits. Duodenal intubation was not deemed necessary in the

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