TOF

:Objectives

Define the TOF and its pathology.1
.Describe clinical manifestations of TOF.2
.Identify physical signs of TOF.3
.Describe management of TOF.4

Tetralogy of Fallot

Prevalence

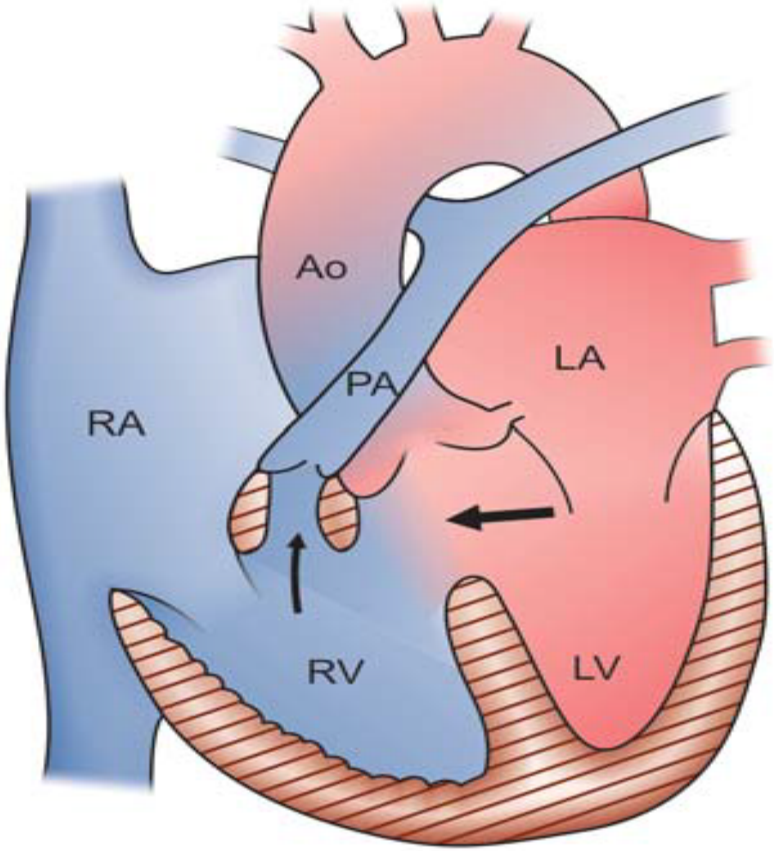
Tetralogy of Fallot occurs in 5% to 10% of all CHDs. This is probably the .most common cyanotic heart defect

Pathology

The original description of TOF included the following four. 1 abnormalities: a large VSD, RVOT obstruction, RVH, and overriding of the aorta. In actuality, only two abnormalities are required, a VSD large enough to equalize pressures in both ventricles and an RVOT obstruction. The RVH is secondary to the RVOT obstruction, and the overriding of the aorta varies

The VSD in TOF is a large perimembranous defect with extension. 2 .into the subpulmonary region

The RVOT obstruction is most frequently in the form of infundibular. 3 stenosis (45%). The obstruction is rarely at the pulmonary valve level .(10%)


Ao
PA
LA
RA
RV
LV

Complete AV canal defect occurs in approximately 2% of patients with TOF, more commonly among patients with Down syndrome, called “canal tet.” In these patients, the VSD has a large outlet component in addition to the inlet portion .associated with the AV canal

Clinical Manifestations

History

.A heart murmur is audible at birth. 1

Most patients are symptomatic with cyanosis at birth or. 2
.shortly thereafter

Dyspnea on exertion, squatting, or hypoxic spells develop later
).even in mildly cyanotic infants

Occasional infants with *acyanotic* TOF may be asymptomatic. 3
or may show signs of CHF from a large left-to-right ventricular
.shunt

)Physical Examination

Varying degrees of cyanosis, tachypnea, and clubbing (in older. 1
.infants and children) are present

RV tap along the left sternal border and a systolic thrill at the. 2
.upper and mid-left sternal borders are commonly present (50%)

An ejection click that originates in the aorta may be audible.. 3

The S2 is usually single because the pulmonary component is too soft to be heard. A long, loud (grade 3 to 5 of 6) ejection-type systolic murmur is heard at the upper left sternal borders. This murmur originates from the PS but may be easily confused with .the holosystolic regurgitant murmur of a VSD

The more severe the obstruction of the RVOT, the shorter and .softer the systolic murmur

Electrocardiography

Right-axis deviation (RAD) (+120 to +150 degrees) is present. 1
.in cyanotic TOF. In the acyanotic form, the QRS axis is normal
RVH is usually present, but the strain pattern is unusual. 2
(because RV pressure is not suprasystemic). BVH may be seen
.in the acyanotic form. RAH is occasionally present

Radiography

Cyanotic Tetralogy of Fallot

The heart size is normal or smaller than normal, and. 1
pulmonary vascular markings are decreased. “Black” lung fields
.are seen in TOF with pulmonary atresia
A concave main PA segment with an upturned apex (i.e.,. 2
.“boot-shaped” heart) is characteristic
RA enlargement (25%) and right aortic arch (25%) may be. 3
.present

Hypoxic Spell

Hypoxic spells (also called cyanotic spells, hypercyanotic spells, “tet” spells) of TOF are not as common as they used to be because many of the patients with TOF receive surgery before they develop the spells. However, it is very important for physicians to be able to immediately recognize and treat the spells appropriately because they can lead to .serious complications of the CNS

Hypoxic spells are characterized by a paroxysm of hyperpnea (i.e., *rapid* and *deep* respiration), irritability and prolonged crying, increasing cyanosis, and decreasing intensity of the heart murmur. Hypoxic spells occur in infants, with a peak incidence between 2 and 4 months of age. These spells usually occur in the morning after crying, feeding. A severe spell may lead to limpness, convulsion, cerebrovascular accident, or even death. There appears to be no relationship between the degree of .cyanosis at rest and the likelihood of having hypoxic spells

.Treatment of the hypoxic spell

.The infant should be picked up and held in a knee–chest position. 1

Morphine sulfate, 0.2 mg/kg administered subcutaneously or. 2
intramuscularly, suppresses the respiratory center and abolishes
.hyperpnea (and thus breaks the vicious cycle)

Oxygen is usually administered, but it has little demonstrable effect. 3
.on arterial oxygen saturation

Acidosis should be treated with sodium bicarbonate ($NaHCO_3$), 1. 4
mEq/kg administered IV. The same dose can be repeated in 10 to 15 minutes. $NaHCO_3$ reduces the respiratory center–stimulating effect of acidosis. With the preceding treatment, the infant usually becomes less cyanotic,and the heart murmur becomes louder, which indicates an increased amount of blood flowing through the stenotic RVOT. If the hypoxic spells do not fully respond to these measures, the following medications can be tried: 1. Ketamine, 1 to 3 mg/kg IV over 60 min. It increases the systemic vascular resistance (SVR) and sedates
.the infant

Propranolol, 0.01 to 0.25 mg/kg (average, 0.05 mg/kg) administered. 2
by slow IV push, reduces the heart rate and may reverse the spell

Management

Medical

1. Physicians should recognize and treat hypoxic spells.
2. Oral propranolol therapy, 0.5 to 1.5 mg/kg every 6 hours, is occasionally used to prevent hypoxic spells while waiting for an optimal time for corrective surgery in the regions where open heart surgical procedures are not well established for small infants.
3. Balloon dilatation of the RVOT and pulmonary valve, although not widely practiced, has been attempted to delay repair for several months.
4. Relative iron-deficiency states should be detected and treated. Iron deficient children are more susceptible to cerebrovascular complications.

Normal hemoglobin or hematocrit values or decreased red blood cell indices indicate an iron-deficiency state in cyanotic patients.

Surgical Palliative Shunt Procedures

Indications: Neonates with TOF and pulmonary atresia

Infants with hypoplastic pulmonary annulus, which requires a. 2
transannular patch for complete repair

.Children with hypoplastic pulmonary artery. 3

Procedures, Complications, and Mortality

Although several procedures were performed in the past
a modified Blalock-Taussig,BT (Gore-Tex interposition) shunt is the
.only procedure performed at this time

Classic BT shunt, anastomosed between the subclavian artery and. 1
the ipsilateral PA, is usually performed for infants older than 3
months because the shunt is often thrombosed in young infants. A
right-sided shunt is performed in patients with left aortic arch; a left-
.sided shunt is performed for right aortic arch

With a modified BT shunt, a Gore-Tex interposition shunt is. 2 placed between the subclavian artery and the ipsilateral PA. This is the most popular procedure for any age, especially for infants younger than 3 months of age. Whereas a left-sided shunt is preferred for patients with left aortic arch, a right-sided shunt is preferred for patients with a right aortic arch. The surgical mortality rate is 1% or less

Complete Repair Surgery

Timing of this operation varies from institution to institution, but .early surgery is generally preferred

Procedure

Total repair of the defect is carried out under cardiopulmonary bypass, circulatory arrest, and hypothermia. The procedure includes patch closure of the VSD, preferably through transatrial and transpulmonary artery approach rather than right ventriculotomy, widening of the RVOT by division or resection of .the infundibular tissue; and pulmonary valvotomy