Eisenmenger's syndrome or Eisenmenger's reaction - 2009

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If we observe bidirectional shunt clearly we are in directing for a picture of significant pulmonary hypertension that accustoms to be observed in the greats ASD of the aged ones that they had not been diagnoses or they had passed unfurnished.
Eisenmenger's syndrome or Eisenmenger's reaction is defined as the process in which a left-to-right shunt (reversed) or bidirectional at the atrioventricular, or aortopulmonary secondary to pulmonary hypertension (pulmonary vascular disease) which in turn, causes increased pressures in the right side of the heart and reversal of the shunt into a right-to-left shunt consequence of high pulmonary vascular resistance of 800 dynes/sec/cm.
Hemodynamic group VI:, reversal flow. Mean frontal QRS axis on ECG 150^0**.**
Eisenmenger's syndrome was so named **Wood, P. *Pulmonary hypertension with special reference to the vasoconstrictive factor.* Br Heart J 1958;20:557-570.** by Dr. Paul Wood after Dr. Victor Eisenmenger, who first described **Eisenmenger V. *Die angeborenen Defekte der Kammerscheidewände des Herzens.* Zeitschr Klin Med 1897; 32(Supplement):1-28** the condition in 1897.

Conditions needed for a person to be diagnosed with Eisenmenger's Syndrome are:

1. An underlying heart defect that allows blood to pass between the left and right sides of the heart. Eisenmenger's syndrome progresses over time as a result of the effects of high blood pressure in the lungs. This high blood pressure, or pulmonary hypertension, occurs because of congenital heart defects that cause blood flow from the left side of the heart to the right side

of the heart (left-to-right shunt). Congenital heart defects of this type include: **1a)** patent ductus arteriosus (PDA) - a connection between the aorta and the pulmonary artery which allows oxygen-rich (red) blood that should go to the body to recirculate through the lungs.**1b)** atrial septal defect (ASD) - an opening in the atrial septum, or dividing wall between the two upper chambers of the heart known as the right and left atria. **1c)** ventricular septal defect (VSD) - an opening in the ventricular septum, or dividing wall between the two lower chambers of the heart known as the right and left ventricles. **1d)** atrioventricular canal defect (AV canal) - a complex heart problem that involves several abnormalities of structures inside the heart, including an ASD, VSD, and improperly formed mitral and/or tricuspid valves. Because the pressures within the left side of the heart are normally greater than those within the right side of the heart, an opening between the left and right side of the heart will cause blood to flow from the left side of the heart into the right side. This left-to-right shunting of blood within the heart causes increased blood flow in the blood vessels of the lungs. Over time, the increased blood flow in the lungs' blood vessels causes increased pressure in these vessels (pulmonary hypertension). If the pulmonary hypertension continues without treatment, the pressure in the right side of the heart may increase to the point that the right side pressure is greater than the left. When this occurs, blood will flow from the right side of the heart to the left (right-to-left shunt), which means that oxygen-poor blood is mixed with the oxygen-rich blood pumped out to the body from the left ventricle.

2. Pulmonary hypertension, or elevated blood pressure in the lungs
3. Polycythemia, an increase in the number of red blood cells
4. The reversal of the shunt.

Etiology A number of congenital heart defects can cause Eisenmenger's syndrome, including atrial septal defects (ASD), ventricular septal defects, patent ductus arteriosus, and more complex types of acyanotic heart disease.

Pathogenesis

The left side of the heart supplies to the whole body, and as a result has higher pressures than the right side, which supplies only deoxygenated blood to the lungs. If a large anatomic defect exists between the sides of the heart, blood will flow from the left side to the right side. This results in high blood flow and pressure traveling through the lungs. The increased pressure causes damage to delicate capillaries, which then are replaced with scar tissue. Scar tissue does not contribute to oxygen transfer, therefore decreasing the useful volume of the pulmonary vasculature. The scar tissue also provides less flexibility than normal lung tissue, causing further increases in blood pressure, and the heart must pump harder to continue supplying the lungs, leading to damage of more capillaries. The reduction in oxygen transfer reduces oxygen saturation in the blood, leading to increased production of red blood cells (minimal polycythemia) in an attempt to bring the oxygen saturation up. Desperate for enough circulating oxygen, the body begins to dump immature red cells into the blood stream. Immature red cells are not as efficient at carrying oxygen as mature red cells, and they are less flexible, less able to easily squeeze through tiny capillaries in the lungs, and so contribute to death of pulmonary capillary beds. The increase in red blood cells also causes hyperviscosity syndrome. A person with Eisenmenger's Syndrome is paradoxically subject to the possibility of both uncontrolled bleeding due to damaged capillaries and high pressure, and random clots due to hyperviscosity and stasis of blood. The rough places in the heart lining at the site of the septal defects/shunts tend to gather platelets and keep them out of circulation, and may be the source of random clots. Due to increased resistance, pulmonary pressures increase sufficiently to cause a reversal of blood flow, so blood begins to travel from the right side of the heart to the left side, and the body is supplied with deoxygenated blood, leading to cyanosis and resultant organ damage.

Physical examination

Any evidence of central cyanosis is frequent.

The face, chest and body are thin, and there may be prominence of the precordium
Clubbing of the finger develops gradually and when present, is usually minimal in childhood,
Cardiac impulse: Right ventricle can be palpated on the left precordial border.
Thrill: eventually present
Heart Murmurs is loud and harsh and extends all through systole, appearing to include both the first and the second sounds in its scope. The point of maximum intensity is between the third and fifth interspaces near the left sternal border.
A pulmonary diastolic murmur of valve insufficiency begins to develop particularly in those with dilated pulmonary arteries and slowly progressive pulmonary disease
Heart sounds: The pulmonary component of the second sound is accentuated.
The pulmonary component of the second sound may be so loud as to overshadow the aortic component. The splitting is not heard in this cases.
A third heart sound in the mitral area is frequent.
A sistolic click may be heard up the left sternal border in some cases of pulmonary hypertension.
The following are the most common symptoms of Eisenmenger's syndrome. However, each child may experience symptoms differently, and each individual may have wide variation of symptoms from mild to severe.

The signs and symptoms of Eisenmenger's syndrome in the advanced stages include:

1) Central cyanosis a blue tinge to the skin resulting from lack of oxygen cyanosis (pale blue or grayish skin due to decreased oxygen in the blood)
2) Dyspnea on exertion (shortness of breath with activity)
3) Fatigue
4) Haemoptysis
5) Palpitations (heart "racing")
6) Dizziness, fainting, called syncope
7) chest pain

8) Right-sided heart failure.
9) High red blood cell count
10) Swollen or clubbed finger tips
11) Arrhythmias
12) blurred vision
13) Bleeding disorders hemorrhage (bleeding)
14) brain abscesses
15) Coughing up blood blood clots (e.g., deep vein thrombosis in extremities)
16) Iron deficiency
17) Kidney problems
18) Stroke
19) Gout
20) Gallstones
21) kidney failure
22) headache
23) dizziness or syncope (fainting)
24) paresthesias (numbness and tingling)
25) stroke
26) gout

ELECTROCARDIOGRAPHIC FEATURES

The typical pattern is right atrial and right ventricular enlargement. Peaked P waves of right atrial enlargement are frequent. A pure right ventricular enlargement pattern is found in an overwhelming proportions of cases.

QRS Axis: Kidd et al. have set six hemodynamic groups in ventricular septal defects. When the mean QRS frontal axis is related to the hemodynamic groups, the findings are as follows:

1) *Hemodynamic group I:* low flow, low resistance: mean frontal QRS axis 30 degree
2) *Hemodynamic group II:* Increased flow, low resistance: mean frontal QRS axis 50 degree
3) *Hemodynamic group III:* Increased flow, slightly resistance: mean frontal QRS axis 70 degree
4) *Hemodynamic group IV:* Good flow, greater resistance mean frontal QRS axis 85 degree

5) *Hemodynamic group V:* Low flow, high resistance mean frontal QRS axis 115 degree

6) *Hemodynamic group VI:* high resistance (800 dynes/sec/ cm^{-5}.), reversal flow. mean frontal QRS axis 150^0.

*Tipically ASD has RV eccentric or volumetric diastolic enlargement pattern: chronic RV volume enlargement causes basically dilatation with proportionate eccentric hypertrophy. The hypertrophied portion of the RV is predominantly the crista supraventricularis (RVOT). The typical pattern of diastolic RVE is incomplete RBBB with rsR' or rsr' patterns in V1. T*all, monophasic R wave broad QRS complexes over the right precordium with tall T waves following them are indicative of marked right ventricular loading, and in such cases the prognosis is poor and almost invariably associated with increased pulmonary vascular resistance. In left precordial leads V5-V6 rS pattern is the rule. There are a small r and a deep S in V5-V6. Prominent R wave are registered in VR lead. Regrettably, this pattern is not exclusive of volumetric RVE, since it may be observed in normal people and in RVE of pressure, such as: moderate PS and mitral valve stenosis with PH.The incidence of IRBBB pattern is of 50% or greater, particularly the younger the group studied, the more frequent the V3R and V4R leads are recorded.The QRS loop of VCG usually is clockwise rotation and located predominantly on anterior and to the right.(Type A VCG RVH). Eisenmenger's syndrome is in the top of spectrum with high resistance and reversal of flow

THE TRIPHASIC PATTERN OF ASD IN V1 OF IRBBB TYPE MAY BE CONDITIONED BY:

I) Right branch congenitally twice longer than normal

II) Selective hypertrophy of crista, responsible for the delay of the final basal vector, which heads to the front and the right

III) Compression or right branch as a neuropraxic phenomenon, in the region of the moderator band.

The RVH type D or IV, has been described by Ramana Reddy, C.V. and Gould, I. A., from the Methodist Hospital from Brooklyn, New York1. These cases would exist in selective RVE of the inferior right paraseptal region of the RV, explored by the V3 and V4 leads. The

ECG shows RBBB. The etiologies are those that evolve with RV volume enlargement and the most representative example is ASD, which causes eccentric dilatation of the chamber with selective predominance of hypertrophy in the RV outflow tract or crista supraventricularis, responsible in part for the Electrovectorcardiographic pattern of ECD (end conduction delay) or IRBBB morphology and triphasic pattern of rsR' pattern in the right precordial leads V3R and V1 and frequent tetraphasic pattern (rsr's') in V3 and V4 translating the selective hypertrophy of the inferior right paraseptal region. More than 95% of the ASDs show this pattern and only 7% normal pattern of the rS type in V1. Vectocardiographically, RV volume enlargement is translated in the HP by RVE of type D or IV, C or III or even B or A. Type A or I (rare) may be found in adults that developed pulmonary hypertension. The vectocardiographic loop of RVE type D in the HP, shows the efferent branch of counterclockwise rotation and the afferent one of clockwise rotation. The initial forces are located in the right anterior quadrant and do not present delay. In isolated RBBB, without RVE, the final forces located in the right anterior quadrant with "glove-finger" shape, present characteristic ECD. The hypertrophied portion of the RV is predominantly the crista or RV outflow tract. It is typically found in ASD (93%). Moderate PS and MS with PH.
ELEMENTS THAT SUGGEST RVE IN V1 IN THE PRESENCE OF RIGHT BUNDLE BRANCH BLOCK (RBBB)
Criteria ofV1 R' wave voltage in Complete RBBB and Incomplete RBBB for RVE.

1) Ramana Reddy CV & Gould LA. Correlative Atlas of Vectorcardiograms and Electrocardiogram. Chapter II pag 43. Futura Publishing Company, INC. Mount Kisco, New York, 1977.