A "four-leaf clover" aortic valve

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A 75-year-old man with acute worsening of heart failure symptoms and a clinical history of prior rheumatic fever, presented with blood pressure 150/60 mmHg and a high-pitched diastolic murmur in the aortic area.

A preliminary Transthoracic exam showed severe aortic regurgitation (vena contracta width >7 mm) (*Figure 1*) and moderate mitral regurgitation due to rheumatic leaflets retraction.

A three-dimensional reconstruction of short axis aortic view showed a four leaflets valve with a significant central tip malcoaptation (*Figure 2, Panel A*).

A Transesophageal exam confirmed a quadricuspid valve with four cusps approximately of equal size (Hurwitz and Roberts Type A) (*Figure 2, Panel B*).

The peculiar spatial arrangement allowed the cusps leading edges to malcoaptate causing a severe aortic regurgitation.

No anomalous origin of coronary ostia was found.

The surgical inspection confirmed the diagnosis (*Figure 3*) and the aortic valve was successfully replaced with a biological prosthesis.

Normally shaped aortic valve has three leaflets according to a regular cono-trunkal embryological ripartition into aortic and pulmonic valve and development of three

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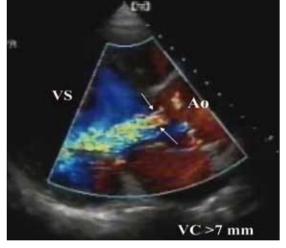


Figure 1 - The Standard Transthoracic approach shows a severe aortic insufficiency (vena contracta width > 7 mm).

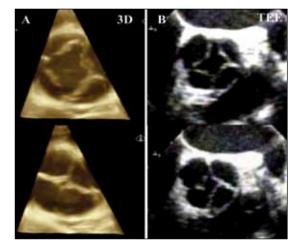


Figure 2 - Panel A. Transthoracic Three-Dimensional echocardiographic reconstruction of the four-leaf clover aortic valve. Panel B. The Standard Transesophageal aortic short axis view shows the symmetrical central tip malcoaptation.

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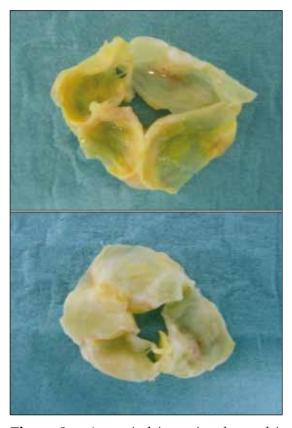


Figure 3 - At surgical inspection the quadricuspid aortic valve appears to be made up of four cusps approximately of equal size. No anomalous coronary ostia were detected.

small pads of mesenchymal tissue. Any eventual deviation from the symmetric arrangement can lead to leaflets number or position abnormalities (1).

According to autopsy series the prevalence of quadricuspid aortic valve ranges between 0,008 and 0,033 % (2) and the valve malformation can be alone or associated to other congenital cardiac defects (3); in 10 % of cases a displacement of coronary ostia or a single coronary artery can be detected (4).

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