

### Flail of the tricuspid valve as a manifestation of neonatal lupus

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**Introduction (or Basis or Objectives):** •Valve dysfunction is not a well-known feature for infants of anti-Ro/SSA-positive pregnancies, but anecdotal cases have suggested the association between rupture of the AV valve tensor apparatus and maternal anti-Ro/SSA antibodies.

**Methods:** •We present a case of a patient with valve dysfunction secondary to rupture of the papillary muscle of tricuspid valve affected by maternal anti-Ro antibodies.

**Results:** •Routine 20 week gestation fetal echocardiography of a first gravida mother revealed focal areas of increased echogenicity at the level of the papillary muscles and chordae of both AV valves (Figure). Heart rhythm was normal. During the following weeks those patchy areas disappeared. Neither insufficiency nor stenosis was detected. Rheumatologic prenatal profile was positive for anti-Ro/SSA antibodies and anti-La/SSB without clinical systemic lupus erythematosus and other connective tissue. At 39 weeks Emergency caesarean section was performed due to fetal hydrops. After birth, the patient required high-frequency ventilation, nitric oxide, inotropic support and diuretic. Postnatal echocardiography showed normal biventricular function and severe tricuspid regurgitation, prolapse of the anterior leaflet with an image that suggested a vegetation (Figure). Laboratory study was positive for factor V Leiden mutation and anti-Ro/SSA antibodies; anti-La/SSB was negative.

Transesophageal echocardiography revealed a flail of the anterior tricuspid valve leaflet. Intraoperative findings showed disruption of the chordal attachments of the anterior leaflet without the presence of a papillary muscle. From the right ventricle a thick bundle muscle of the free wall was detached and used as a neo-papillary muscle. The anterior leaflet of the tricuspid valve was directly attached to the neo-papillary muscle with prolene 5/0. A tricuspid annuloplasty was also realized (Figure). Biopsy specimens from the right ventricle were obtained.

**Conclusions:** The pathogenesis of the papillary muscle chordal rupture may be similar to that proposed for autoimmune AV block, binding in this case the antibodies to the cardiac myocytes and evoking an inflammatory response with subsequent fibrosis and ultimately muscle chordal rupture.

**Figure:** Fetal echocardiogram (a) with localized areas of increased echogenicity at the papillary muscles of both AV valves. Postnatal echocardiogram showed severe insufficiency of the tricuspid valve and an image that reminded a vegetation (b). Echocardiogram showed a flail of the tricuspid valve with severe regurgitation and dilatation of the right ventricular cavities (c). Postoperative results with good leaflet coaptation and mild regurgitation (d).

