Material & Methods:
From the hospital database/chart, data were obtained. Echocardiogram studies were reviewed and correlation with anatomical features from pathological study was done.

Four cases were studied with absence of left AV connection with a dominant left ventricle and rudimentary right ventricular chamber located at the right side. Three specimens were examined from patients who died. The remaining patient has been followed after a palliative surgery.

Results:
There was agreement between the initial echocardiogram report and pathological features in two cases. In the two remaining, the initial diagnosis was "Tricuspid atresia", including one fetal diagnosis.

In all cases there was an early clinical presentation with hypoxia and low cardiac output. Right outflow tract obstruction was detected in two patients. Two patients required atrial septostomy for a restrictive PFO. In the remaining patient there was a small PFO, but there was total anomalous pulmonary venous return to the coronary sinus.

Main echocardiographic features:

**Patient** | **ASD** | **VSD** | **Ventriculo-arterial connections** | **Location of rudimentary chamber** | **RVOTO** | **TAPVD**
---|---|---|---|---|---|---
1 | PFO | Muscular restrictive | concordant | Right | yes | No
2 | Restrictive PFO | Muscular restrictive | discordant | Right | No | No
3 | Secundum ASD | Non restrictive muscular | concordant | Right | No | No
4 | PFO | Non restrictive muscular | concordant | Right | yes | Yes

Conclusions:
Echocardiographers must be aware about unusual forms of AV connection. The correct interpretation of absent AV connection by standard views needs a dynamic evaluation and the combination of different views to establish the correct spatial orientation of cardiac chambers and determination of ventricular morphology. Functional diagnosis requires more views and careful interpretation as well. The sequential segmental analysis is fundamental to achieve the correct diagnosis, both by echocardiography and other morphological methods.
Despite of the excellent outcome of AVSD repair achieved in the last 50 years, the left AVV remains the “achilles” heel for surgeons. Several surgical techniques has been used, focusing in minimizing residual AVV regurgitation. However, there is a paucity of data regarding of left AVV stenosis. We sought to identify the incidence, late outcome and risk factors related with this morbidity.

Material & Methods:
221 patients underwent to all types of AVSD repair. Routine intraoperative transesophageal echocardiogram (TEE) were performed in all above 3.5 kg. Among them 18 presented residual Left AVV stenosis (mean gradient ≥ 8 mmhg) (group I). Age at the surgery (4 months to 22.5 years/median of 11 month). Group II comprised 26 patients with trivial/mild residual LAVV regurgitation. Measurements of AVV annulus, distance between the two left papillary muscle were obtained from the pre and post-operative TEE.

Clinical and surgical data were obtained by chart review.

Results:
There were 6 re-operations, including 2 valve replacement and 3 deaths in the early post-op. No late post-op death. Two patients lost follow-up. Among 13 remaining, there was one late re-operation. The LAVV gradient remained stable and/or lower over the time. Mean follow-up was 6.6 years (0.8 to 10.7 y).

Preoperative total common valve annulus and LAVV annulus, measured by intraoperative TEE, were not different between groups. However, the preoperative distance between the papillary muscles were significantly smaller in Group I (p = .01). (graphic 1)

Postoperatively, LAVV annulus were reduced comparing with preoperative values, in Group I (p < .001), but not in Group II. (graphic 2)

Pre-op moderate to severe LAVV regurgitation correlated with the occurrence of residual LAVV stenosis (table 1).

Partial AVSD, Down’s Syndrome, Tetralogy of Fallot and Heterotaxy syndrome were not risk fact, as the surgical technique neither.

Conclusions:
Residual LAVV stenosis occurred in 8.1%.

The distance between the two left papillary muscles (which support the trileaflet LAVV and also predict the mural leaflet length) was significantly smaller in those patients with residual LAVV stenosis.

The LAVV annulus decreased after repair in patients whose LAVV became stenotic. The need for further re-operation was low and overall patients remained stable from the clinical point of view.