**Objectives**

The goal of this study was to evaluate the associated conditions and outcome of fetuses with agenesis of ductus venosus (ADV).

**Methods**

Retrospective study of the ADV cases evaluated during a five-year period in a referral tertiary unit, detected during the first and second trimester screening and referral scans. We evaluated the prevalence, umbilical shunting characteristics, concomitant anomalies, and outcome.

**Results**

We identified 9 cases of ADV (1 in 388). Major structural defects and fetal effusions were detected in 66% and 55% had a major chromosomal abnormality.

In 33% ADV was apparently isolated and two of three (with normal portal system development) had a normal outcome at one year after birth, while the third (with abnormal portal system) presented digestive anomalies and growth retard after birth.

In our small series the outcome of fetuses with ADV was independent of liver by-pass, caliber of the shunt, and NT thickness, but influenced by the presence of associated malformations and portal development.

We detected two new types of umbilical drainage (complex: intra- and extrahepatic and double: cardiac concomitant with normal, through ductus venosus).

**Conclusions**

There is a wide variation in reported ADV incidence that depends on the population risk. The detection is important, given the high association with major abnormalities. Associated malformations, including abnormal portal system development represents key predictors for outcome.