

## Embryology of arrhythmogenic right ventricular dysplasia (ARVD)

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**A** case of arrhythmogenic right ventricular dysplasia (ARVD) was discovered during routine echosonography at 24 weeks of gestation. The four-chamber view showed a large aneurysmatic area extending from below the tricuspid valve to the insertion of the moderator band; the affected wall appeared thin and akinetic, with no flow at color Doppler investigation, and no evidence of cardiovascular failure. The size of the aneurysmatic area was unchanged at subsequent controls (25 and 26 weeks of gestation). Arrhythmias could be ventricular or atrial since the involvement of the atrium is frequent. The pregnancy ended in spontaneous abortion at 27 weeks. The histopathologic examination of the heart showed the presence of adipocytes interspersed with myocardial fibers, confirming the diagnosis of ARVD. The zone of highest amount of adipocytes was located

in the mediomural layers confirming where the disease starts in the embryo. This is logical because anomalies in desmosomes is the most frequent genetic factor. As the right ventricle is made of two perpendicular layers it is possible to suspect that during embryogenesis a shearing effect is taking place between the two layers especially because qtg that time the right ventricle was systemically generating strong biomechanical forces. Subsequently the sub epicardial layers were affected more severely than the subendocardial layers. When the subepicardial layers were almost completely destroyed the disease seemed to start from the epicardium towards the endocardium. However, some remnants could be visible in most of the cases if this is observed carefully. This case remains unique in the literature.

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