Long-term results of heart transplantation (HTX) in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC)

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is associated with myocardial fibrosis, fibrofatty replacement, and a progressive loss of predominantly right ventricular tissue, although biventricular disease involvement is not uncommon. Ventricular arrhythmias may cause sudden cardiac death (SCD) in ARVC patients. Disease causing mutations in genes encoding for desmosomal proteins have been reported in more than 40–50% of ARVC patients.[1,2]

To evaluate the correct timepoint for heart transplantation (HTX) remains a difficult and crucial task. As left ventricular function is maintained for a long time in normal ranges and onset of arrhythmogenic complications can be unpredictable, patients with ARVC are threatened by rhythmogenic death prior to HTX.

We report of our institutional experience in ARVC pts. since 03/1989.

18 pts. with ARVC (10 male/8 female) (Age range 16–69 years/mean: 45 yrs.) have been transplanted until 06/2014. 14 of 18 pts. have been provided with either ICD (12) or BV-ICD (2) support prior to HTX. 14 pts. needed inotropic support, 2 pts. additional support by intra-aortic balloon pump (IABP). 2 pts. had to be supported by mechanical assist devices.16 pts. had to be listed as „high urgent“ pts. (adequate to UNOS 1a) with a waiting time of 3-230 days/mean: 53 days). 2 pts., waiting 7 and 1363 days have been transplanted from the elective list. 2 pts. with VAD support aquired waiting times of 34 and 230 days. None of the pts. had secondary pulmonary hypertension, even after longer waiting times (last PVR prior to HTX 61-231 dyn*sec*cm^{-5}/mean: 142 dyn*sec*cm^{-5}). At time of analysis 61,1% of pts. are still alive after HTX with survival times of 1,4 to 211,6 months (mean: 61,5 mths.). Causes of death have been rejection, neoplasia, infection, pulmonary, abdominal, renal failure and unknown7lost to follow up in 1 case (5,6%).

For ARVC pts. with the ongoing threat of arrhythmogenic death primary HTX ist he recommended therapeutic option as VAD support in this patient group is difficult and rather experimental. Results after HTX are promising even for younger pts. with ARVC. Decision making for the correct HTX timepoint is difficult and not always transferrable into practice under the circumstances of the German allocation policy.

Due to long waiting times the majority of pts. needed inotropic or mechanical circulatory support prior to HTX.

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