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## Surgical results of Ebstein anomaly based on pathology and techniques

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**Objective:** To evaluate the outcomes of patients who underwent the different techniques according to the pathology of Ebstein Anomaly (EA).

**Method:** From March 2004 to February 2017, 228 patients (mean age, 19.8±15.4 years; range, 7 months-64 years) with EA underwent 232 cardiac operations at our hospital. 29 patients had a prior cardiac procedure before. Among the patients in first procedures, 92 patients (46.2%) were categorized to Carpentier type C and 63 patients (31.7%) were type D, 7 patients (3.5%) had isolated anterior leaflet downward displacement. Anatomical repair were performed in 179 patients (Type B, n=35; type C, n=92; Type D, n=29; unclassifiable, n=7; reoperation, n=16), 1½ ventricle repair in 37 (Type D, n=33; reoperation, n=4), tricuspid valve repair in 3 (Type A, n=2; Type B, n=1), tricuspid valve replacement in 10 (7 reoperations), and Fontan procedure in 3 (TCPC, n=2; Glenn, n=1). Atrialized right ventricle presented in 194 cases (168 excised, 6 incorporated). The pathology of 199 patients who underwent first procedures at our center described in Table 1.

**Result:** The mortality was 1.7% (n=4: anatomical repair, n=3; 1½ ventricle repair, n=1). Among these, 1 pathology type was Carpentier type C and 3 were type D. 1 A-V block (0.4%) newly occurred. 214 patients were available to follow up. The range of follow-up duration was 10 months to 13 years (mean, 7.3±3.2 years). Late survival was 99% (2 late deaths) at 10 years. 3 patients received reoperation (reoperation rate, 1.3%; TVR, n=1; 1½ ventricle repair, n=2). Mean New York Heart Association class improved from 3.5 to 1.1.

**Conclusion:** The principle of the techniques is to reconstruct the tricuspid valve and right ventricle anatomically. For most cases, the anatomical repair was demonstrated with low mortality, less complications and excellent durability at long-term follow-up. The Carpentier classification cannot categorize all the patients. It is critical to choose applicable surgical techniques individually according to the pathologic morphology for EA surgical results. If the tricuspid valve is severely hypoplastic, 1½ ventricle repair and valve replacement may be alternative.

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