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Surgery of single ventricles: is the humanitarian action justified?

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Objectives:
Children with a single ventricle usually require several surgeries before eventually benefiting from a total cavo-pulmonary connection. The aim of this study is to know if such support is possible for children from developing countries, managed by an humanitarian association, taking in account the follow-up and the survival improvement, and if so, for what types of single ventricles.

Methods:
We studied retrospectively all single ventricle patients managed by our association Mécénat-Chirurgie Cardiaque (MCC) since its creation in 1996. After their return to their country, the children were followed up in collaboration with the corresponding doctors in the countries of origin.

Results:
From 1996 to 2017, 138 children (90 boys, 48 girls) with single ventricle were managed by MCC at a mean age of 4 years. 33% are from sub-Saharan Africa, 25% from North Africa, 24% from the Middle East and 15% from Eastern Europe. 19 children were withdrawn from surgery because of pulmonary hypertension or overly complex heart disease. 119 were operated on, with a total of 165 procedures: 41 had only palliative surgery (systemic-pulmonary anastomosis or banding), 47 a partial cavo-pulmonary connection (PCPC), and 31 a total cavo-pulmonary connection (TCPC). The mean age at the TCPC was 8.5 years. The overall operative mortality was 10/160 (6.2%). After a mean follow-up of 5.5 years, 18 children (13%) were lost to follow-up. Survival rate of operated children was 82% and 79% at 5 and 10 years, compared with 39% and 29% for non-operated children. The prognosis is better for tricuspid atresia (90 and 86%) and double inlet ventricles (87 and 83%) than for DORV or atrio-ventricular canal (64% and 68% at 5y).
After palliative surgery, survival rate was 72% at 5y and 63% at 10, whereas after PCPC it was 81% and 77% respectively, and 97% 10 years after TCPC.

Conclusion:
Surgical management of children with single ventricles from developing countries by a humanitarian association is possible and legitimate, with a very satisfactory long-term follow-up and survival rate. Favourable forms are tricuspid atresia or double inlet single ventricles. The management should aim towards a TCPC as soon as possible.