Late management of truncus arteriosus: 20 years of humanitarian experience

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Background: the spontaneous evolution of the truncus arteriosus is poor because of the earliness of PAH. In developed countries, children are operated in the neonatal period with excellent long-term results. Late diagnosis and management remain rare, except in developing countries.

Aim: to describe the prognosis of children with truncus arteriosus diagnosed and tardily managed in developing countries.

Results: for 20 years, the humanitarian organization Mécénat-Chirurgie Cardiaque (France) has supported 41 children with truncus arteriosus from developing countries. The average age at management was 3 years old. Late diagnosis and management can be explained by the lack of adequate facilities in the developing countries. 8 children without shunt symptoms had to be recused, and 33 have been operated (32 repair surgery, 1 pulmonary banding). Surgery was decided if clinical and radiological symptoms of shunt were persistent, particularly if the cutaneous oxygen saturation was above 88%, regardless of hemodynamic calculation of pulmonary vascular resistance, which was not carried out systematically. The postoperative course was marked by PAH crisis requiring NO ventilation and sildenafil in 30% of cases. The operative mortality has been 1/33. The median stay in ICU was 5 days. The 32 children were able to return home without treatment and are followed up by the corresponding cardiologists, in relation with the French team of MCC. None of the children were lost to follow-up, with a mean follow-up of 3.4 years after surgery. At last follow-up, 1 child died six months after his surgery (infective endocarditis on the tube), 1 child had a massive truncal valve insufficiency, 5 had a significant stenosis of the RV-PA tube (3 symptomatic children are waiting for tube change), and 2 have already been reoperated 4 and 6 years after the complete repair to change the tube.

Conclusion: Late management of truncus arteriosus (even after two years of age) is possible with good long-term results and surgery remain possible without prior hemodynamic examination up to an advanced childhood if persist signs of left-to-right shunt, in particular a saturation above 88 %.