

Original Article

Late management of truncus arteriosus: 20 years of humanitarian experience

Marielle Gouton,^{1,2} Vincent Lucet,^{1,3} Olivier Bical,¹ Francine Leca¹

¹Mécénat Chirurgie Cardiaque, Enfants du Monde; ²Cardiologie Congénitale Montsouris, Institut Mutualiste Montsouris, Paris; ³Centre de Cardiologie Infantile, Le Château des Côtes, Les Loges en Josas, France

Abstract Objectives: Early surgical management of common arterial trunk is well established and has good prognosis. Late diagnosis is less common. We reviewed late-diagnosed common arterial trunk management and prognosis for children in developing countries. We also discuss the need for prior catheterisation. **Material and methods:** We reviewed all common arterial trunk patients managed by our humanitarian organization since 1996. **Results:** A total of 41 children with common arterial trunk were managed at a mean age of 3 years old. The lack of adequate facilities in developing countries explains the late management. The decision to proceed with surgery was based on clinical and radiological symptoms of persistent shunt, particularly a high cutaneous saturation level, regardless of catheterization – not carried out systematically. Eight children had to be withdrawn and 33 (80.5%) received operation – mean saturation 91%. The postoperative course was marked by pulmonary arterial hypertension requiring specific treatment in 30% of cases. The operative mortality was 1/33. The 32 children returned home without treatment after a mean post operative stay of 49 days and were followed up (mean FU 3.4 years, none lost to follow-up). At last contact, 1 child died six months after surgery, 1 child had a massive truncal valve insufficiency, 5 had a significant stenosis of the RV-PA tube, and 2 have had further surgery for tube replacement. **Conclusions:** Late management and surgery of common arterial trunk is possible with good long-term results without prior hemodynamic examination up to an advanced childhood when signs of left-to-right shunt persist. A high saturation level (above 88%) seems to be a good operability criterion.

Keywords: Truncus arteriosus; late surgery; humanitarian

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THE COMMON ARTERIAL TRUNK OR *TRUNCUS arteriosus* IS A RARE CHD (2–3% OF ALL CHD). In the absence of an adapted treatment, its natural prognosis is poor: more than 80% die within the first year of life. This is primarily due to left-to-right shunt-induced cardiac failure. Beyond the first year of life, mortality is associated with the development of precociously irreversible^{1–3} pulmonary arterial hypertension.

The current treatment includes early screening (foetal or neonatal) followed by early surgical repair, around 1 month of age where possible. The purpose of the repair is to close the ventricular septal defect

and to reconstitute the pulmonary outflow tract with a tube, valved or not,^{4,5} between the right ventricle and the pulmonary artery.

Some children miss this early support,^{6,7} either because of lack of diagnosis or because surgical treatment is not available. This occurs in many developing countries. Humanitarian associations can be asked to support those undiagnosed or untreated children with a severe heart who have survived thus far. The association Mécénat Chirurgie Cardiaque (MCC)⁸ is regularly called upon to deal with these children. MCC is a humanitarian association created in 1996. Its mission is to enable children suffering from cardiac malformations to come to France to access the care that is not available in their native countries.

The purpose of this retrospective study was to establish whether children with “late-diagnosed common

Correspondence to: M. Gouton, Mécénat Chirurgie Cardiaque, Enfants du Monde, 33 rue Saint-Augustin, 75002 Paris, France. Tel: +33 6 17 27 16 19; E-mail: m.gouton@cardiocongenitale.fr

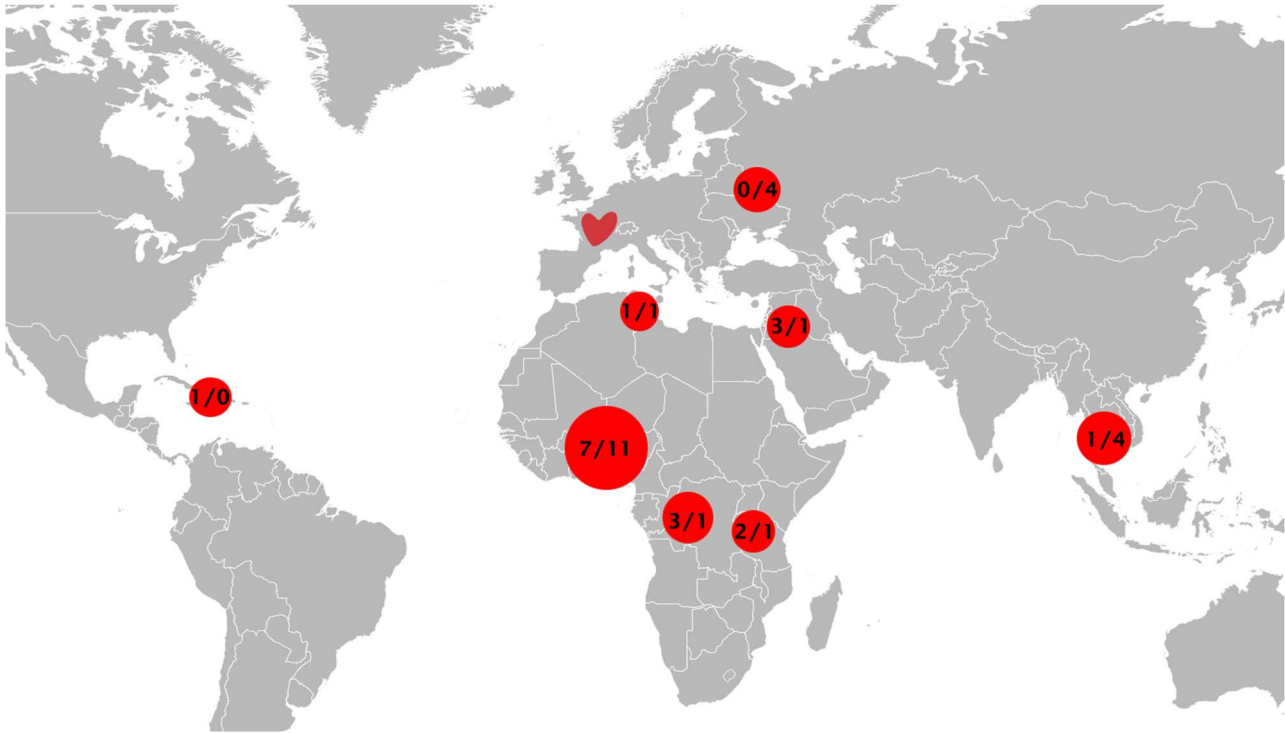


Figure 1.

Countries of origin of the children (number living in the capital/number living in rural areas).

arterial trunk” can benefit from late surgery. Late surgery is defined as beyond the usual criteria of operability of common arterial trunk as described in the literature.^{9–11} We also described short- and mid-term follow-up of these children receiving late operations, and discussed the need for prior catheterisation in this population.

Materials and methods

Study design and population

We reviewed all relevant files – children with common arterial trunk – addressed to MCC since its creation in 1996. The common arterial trunk diagnosis was made by our corresponding cardiologists in their countries of origin. The children’s potential suitability for treatment – and therefore their coming to France – was decided following correspondence between MCC and the corresponding cardiologists, and was based on clinical and radiological signs of shunt.

Since 1996, 41 children were entrusted to MCC for surgical treatment of common arterial trunk (23 boys, 18 girls). The children came mainly from sub-Saharan Africa (26/41–63%), but also from the Middle East and North Africa (5/41–12%), from South-East Asia (5/41–12%), Eastern Europe (4/41–10%), and Haiti (1/41–2%). More than the half of the children (23/41) came from rural locations, while 18 children were living in their country capital (Fig 1).

On arrival in France, each child had a hospital assessment in one of the French centres for congenital heart surgery, which included the following:

- A clinical evaluation in search of persistent signs of a left-to-right shunt – dyspnoea, intercostal retraction, sweating, thoracic deformation, hepatomegaly, insufficient weight gain, insufficient food intake.
- A thoracic radiography in search of a cardiomegaly or of a pulmonary hypervascularisation.
- The most reliable measurement of cutaneous oxygen saturation.
- An echocardiography with Doppler with two expert reviews for each child.
- If needed, a haemodynamic evaluation was carried out, but not systematically, and left to the discretion of each centre.

This evaluation confirmed the diagnosis and the type of common arterial trunk, and decided the operability of the child.

Preoperative data and immediate postoperative data were collected retrospectively in our centralised database based on medical records. Late follow-up was ensured by telephone or e-mail by the responsible for the medical follow-up in our association with our correspondents in the countries – cardiologists or paramedics- or the families of the children.

Statistical analysis

Statistical analysis was performed with Stata10.1 software (StataCorp LLC, College Station, Texas, United States of America). Qualitative data were described with sample sizes and percentages. Numeric quantitative data were calculated by mean, median, and standard deviation. Rates were compared with the χ^2 test or exact Fisher test if sample size was <5. Means were compared with the Student's t-test. Survival was analysed by the Kaplan–Meier actuarial method. The p-value was statistically significant if ≤ 0.05 .

Results

Initial characteristics

The characteristics of the population are detailed in Table 1.

In all the 41 cases, the common arterial trunk was type I–II of the Van Praagh classification.¹² In addition, four patients had a moderate pulmonary stenosis, two patients a coronary anomaly, single coronary artery, and one patient had mitral stenosis. The truncal valve was quadricuspid in nearly 40% of the cases, with a valve insufficiency grade I (16/41), grade II (9/41), or grade III (5/41), and a tight stenosis (2/41).

The children's management changed over the 20 years of our study, especially the performance of a haemodynamic examination that was more often performed in the early period, whereas the characteristics did not change with time (Table 2).

Operability and surgery

In total, 20 children underwent catheterisation; six of them were withdrawn from surgery thereafter because of a high level of indexed pulmonary vascular resistances (15.8 WU) and pulmonary/systemic resistance ratio (0.69) and no O₂ or NO pulmonary vasoreactivity. Two children were withdrawn without catheterisation because of the absence of any clinical shunt sign, absence of radiological shunt (cardio thoracic index (CT)=0.55), and a low saturation (76 and 82%, respectively). Among the eight withdrawn children, only one was withdrawn in the recent period (cardio-thoracic index 0.55, saturation 76%, no catheterisation).

Finally, 33 children out of the 41 (80.5%) were operated on (Table 3); they had a median age of 2 years 11 months (5 months – 11 years). They differed from the non-surgical children by their saturation (90 versus 84%; $p = 0.002$) and their CT index (0.62 versus 0.56; $p = 0.007$).

Table 1. Initial characteristics.

	Catheterisation			Recused			Surgery			Total		
	Total (mean)	No catheterisation (mean)	P	Total (mean)	After catheterisation (mean)	Without catheterisation (mean)	P	After catheterisation (mean)	Without catheterisation (mean)	Total recused (mean)	Total surgery (mean)	P
n	41	21		6	14	2		6	2	8	33	
Age (years)	2.94	2.29	0.13	2.66	4.02	2.65	0.64	2.25	2.25	2.66	3.01	0.75
CT index	0.61	0.60	0.18	0.57	0.63	0.55	0.25	0.61	0.61	0.56	0.62	0.007
Saturation (%)	89.6	89.1	0.6	86	92	79	0.25	90	90	84.4	90.9	0.002
Systolic PP				77.6	82.7							0.67
Diastolic PP				61.4	57.4							0.62
Mean PP				42.7	38.4							0.69
QP/QS				1.14	5.4							0.01
iPR (Wood units)				15.8	10.3							0.04
PR/SR				0.69	0.24							0.0007
O ₂ or NO vasoreactivity				0/3	4/5							

CT index = cardio-thoracic index; iPR = indexed pulmonary resistances; NO = nitric oxide; PP = pulmonary pressure; PR/SR = pulmonary/systemic resistances ratio; QP/QS = pulmonary/systemic flow ratio $p < 0.05$ are statistically significant and bold emphasised

Table 2. Change of management with time.

Period (years)	n	Age (years) (mean)	Saturation (%) (mean)	CT index (mean)	Recused without KT (n)	Recused after KT (n)	Surgery after KT (n)	Total KT (n)	Surgery without KT (n)	Total surgery (n)
Until 2003	10	2.9	88.6	0.62	0	2	5	7	3	8
2004 → 2011	10	3.7	89.5	0.59	1	4	2	6	3	5
2012 → 2014	11	2.7	89.9	0.62	1	0	6	6	4	10
Since 2015	10	2.4	90.2	0.62	0	0	1	1	9	10
Total	41				2	6	14	20	19	33

CT index = cardio-thoracic index; KT = prior catheterisation

Table 3. Age of the population.

	Recused	Surgery	Total (%)
<1 year	1	7	8 (19.5)
1–2 years	4	12	16 (39.0)
>2 years	3	14	17 (41.5)
Total	8	33	41

Previously catheterisation was carried out when a doubt about operability persisted, essentially at an advanced age: the 14 who underwent catheterisation were 4 years old compared with 2.2 years in the 19 without catheterisation ($p = 0.04$), but did not differ by their saturation (90% versus 92; ns) or their CT index (0.61 versus 0.63; ns).

A child underwent only a palliative intervention with banding of the pulmonary arteries. All the others benefited from a complete surgical cure – preceded in two cases by a banding – with interposition of a RV-PA tube (Hancock[®] 17 (Hancock, Medtronic, Minneapolis, Minnesota, United States of America), Contegra[®] 7 (Contegra, Medtronic, Minneapolis, Minnesota, United States of America), Homograft 4, Venpro[®] 2 (Venpro, Venpro Corp, Irvine, CA, United States of America), Labcor[®] 2 (Labcor, Labcor company, Brasil)) of 15.6 mm mean diameter (12–21) in all the cases. The choice of the type of tube was left to the surgeon's discretion.

Postoperative course

The immediate postoperative pulmonary arterial hypertension required an adapted pharmacological management, justifying the prescription by the local physician of sildenafil after inhaled NO in more than one-third of the cases. Three children required a surgical recovery: tamponade at the 6th postoperative day, sternitis at day 15, and mediastinitis at day 20. The median stay in intensive care unit was 5 days (2–47, mean 7.7). Eight children required an extended stay in the ICU, prolonged beyond 1 week, primarily because of pulmonary arterial hypertension crisis (4/8), pulmonary infection (2/8), a regressive

coronary ischaemia on a single coronary artery (1/8), or a secondary pericardial drainage (1/8).

The hospital mortality of our series was 1/33: one 7-year-old child died after 2 days of circulatory assistance. This child had no preoperative clinical sign of heart failure, a cutaneous saturation of 89%, a tight stenosis of the truncal valve, but had a cardiomegaly (CT index: 0.68). This child had been operated on without preliminary haemodynamic examination and received a RV-PA Venpro[®] tube and a mechanical Sorin[®] aortic valve.

The pulmonary hypertension evolution was quickly favourable after the surgery, with a postoperative mean systolic right ventricle pressure in the recovery room, estimated at echography with the RV-RA velocity or the left ventricle–right ventricle velocity through a residual ventricular septal defect, at 53 mmHg (83 mmHg preoperative) and 36 mmHg at last follow-up, after a mean postoperative stay in France of 49 days. Sildenafil, when prescribed in the immediate postoperative course, could be stopped in all cases before the return to the child's country, without recurrence of pulmonary arterial hypertension, and without new onset of heart failure.

The mean cost for each stay in France was €12,225 (€396–€41,982).

Follow-up

Among the eight children who had not been operated on, two were lost to follow-up after their return to their country. For the six others, the mean follow-up was 3.5 years (7 months–7 years 10 months), with one death at 8 years old, that is to say, 6 years after being identified as not suitable for surgery. At 1- and 5-year follow-up, survival rate was 100% and 50%, respectively.

All the 32 children who were operated on and returned to their countries were followed up. We have not had any news for more than 5 years regarding four of them (they were followed up at 2.3, 2.6, 4.8, and 9.4 years after surgery). The mean follow-up since surgery was 3.4 years (1.5 months–16 years). We deplore a death 6 months after surgery due to an

infectious endocarditis on the RV-PA Contegra[®] tube in the child who was re-operated on for a stenosis. At 1-, 5-, and 10-year follow-up, survival rate was 93%.

The evolution of the pulmonary arterial hypertension is not known. Nevertheless, clinical outcomes of the children did not differ between those having had Sildenafil in the postoperative course and those who did not. The evolution of the aortic/truncal valve is known for 15 children out of the 31 survivors: one notes an aggravation of the aortic insufficiency in only one patient – from grade I aortic insufficiency at preoperative assessment to grade III at the last control, 11 years after initial surgical repair, waiting for another operation. The other children kept a mild aortic insufficiency (grade 1 or 2), without pejorative evolution during the entire follow-up period. At last control, a mild stenosis of the RV-PA tube (gradient < 40 mmHg) occurs once, a moderate to severe stenosis (gradient > 40 mmHg) five times, including three children who are waiting for a tube change. Two patients have had to be operated on again for the change of their RV-PA tube, 4 and 6 years after the initial repair – change of a Hancock[®] 12 and a Contegra[®] 12, respectively, for a Hancock[®] 16. At the time of the last contact, all children but four – who were waiting for their tube change or aortic valve replacement – were in good health, without treatment.

Discussion

The low mean cost for each child, well below the average cost of cardiac paediatric surgery in French public hospitals, was made possible thanks to the sustained effort of all the people involved in our association – volunteers travelling with children from and back to their country, volunteer host families caring for the children throughout their stay in France, and the medical and paramedical teams of the various hospitals involved –, and thanks to the fixed day price we recently set up with most of the hospitals concerned. All the efforts converge towards a reduced length of stay, with the preoperative assessment carried out on an outpatient basis, and catheterisation – if necessary – carried out on the same day as the surgery with a hospitalisation in the morning and an early postoperative discharge to a paediatric cardiac convalescence care setting. Therefore, no cost consideration interferes in our indication for the haemodynamic examination in these children: if we decide to operate on the child, the hospital cost will be the same with or without prior catheterisation. This organisation allows the management of complex cardiac diseases such as common arterial trunk at a cost acceptable to our association.

If the early surgical management of common arterial trunk is not discussed any more, the invasive preoperative assessment for operability of “out-of-date” cases has to be discussed in the light of the recent imaging techniques – MRI, CT – and of the gained experience.¹³ Some teams remain in favour of the systematic preoperative haemodynamic catheterisation, in particular in children older than 1 year.^{14,15} For others, on the other hand, the catheterisation with calculation of pulmonary vascular resistances does not have preoperative benefit when obvious clinical signs of a significant left-to-right shunt persist and does not have to be systematically done any more, even after 1 year of age. In our series, 78% of the treated children (26/33) were more than 1 year old at the point of surgery, and only 14 out of 26 had a preoperative exploratory catheterisation. Catheterisation was performed less often in the last years, because of a change in our global management. We saw that, independently of their pulmonary vascular resistances measured in cathlab, children with obvious persistent clinical and radiological shunt signs have had a good surgical prognosis, even when needing NO or Sildenafil in postoperative care. Therefore, we decided to operate on children only on clinical, echographical, and radiological residual shunt signs, and to perform catheterisation only in children in whom a doubt of high pulmonary vascular resistance without vasoreactivity persists. Our series thus pleads in favour of a surgical attitude without systematic prior catheterisation, in the classical type 1 and 2 forms of common arterial trunk without associated anomalies, and under conditions of clinical and radiographic signs of left-to-right shunt, in particular with a high peripheral saturation. Because our two populations, operated or non-operated, differ clinically only about cutaneous saturation level (90 versus 84%, respectively; $p = 0.002$), we propose a cut-off between these two means, that is to say, $\geq 88\%$.

The immediate prognosis of the surgery depends on the crisis of pulmonary arterial hypertension, which can require an extended stay in the recovery room, but which evolves generally favourably thanks to the new pulmonary arterial hypertension management, inhaled NO, relayed by sildenafil: only 1 death out of 33 in our series. At 1 year, global mortality in the operated on population is low at 2/33 (6%) when it is greater when surgery is performed earlier in the infancy: 11% between 2005 and 2014 in the series of Naimo et al¹⁶ and 12% in that of Chen et al.¹⁷ The mid- and long-term prognosis of operated common arterial trunk remains marked by the need for another operation either on the truncal valve, or on the pulmonary conduit.¹⁸ In our series, with a mean follow-up of 3.4 years (up to 16 years), we have

not had as yet to operate again on the truncal valve, with a good stability of the aortic insufficiency in time – even if several teams mention good performances after additional operation for severe aortic insufficiency.^{19–21} Nevertheless, the development of a stenosis on the reconstituted pulmonary tract appears unavoidable, and needs later surgeries for RV-PA tube change, roughly 3–4 years after the first intervention.^{22,23} Because of the small size of our series and the multiplicity of the implanted tubes, we cannot conclude as to the superiority of one or the other type of tube on morbidity or mortality.

The follow-up process operated by MCC is strong. Despite the geographical distances involved, we have had no news for over 5 years after initial follow-up for only four of the 41 children concerned. This strong follow-up process allows us to confidently consider managing tube change as the child grows, as well possible new surgery on the aortic valve when the clinical state of the child justifies it.

Conclusion

The mid-term prognosis of late diagnosis and treatment of common arterial trunk (beyond 1 year) appears to be excellent. The surgery criteria are simple: persistence, whatever the age of the child, of clinical and radiological signs of important left-to-right shunt, in association with a high level of cutaneous saturation (cut-off that we defined, with our data, above 88%), without necessarily preliminary catheterisation. In the case of an echographic doubt on associated anatomical anomalies (in particular a pulmonary branch stenosis or hypoplasia), other pre-operative investigations must be made – catheterisation and/or thoracic CT.

The immediate postoperative period is often marked by pulmonary arterial hypertension crisis justifying an optimal postoperative medical management. The long-term prognosis, and the need for re-operation for the RV-PA tube change, which is unavoidable because of the growth of the child, depends on the quality of the long-term follow-up. The close follow-up set up between MCC and the corresponding cardiologists in the origin countries of the children make it possible to be optimistic as for this prognosis.

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Conflicts of Interest

None.

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