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Original Article

A multinational and multidisciplinary approach to treat CHD in paediatric age in Angola: initial experience of a medical-surgical centre for children with heart disease in Angola

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Abstract *Background:* Epidemiological patterns of cardiac disease differ between developed countries and African nations. Despite the collaborative efforts of developed countries, several obstacles hinder the implementation of successful programmes for the management of children with heart disease in Africa. *Materials and methods:* This study is a retrospective analysis of a bi-national two-institution partnership programme for the treatment of children with congenital and acquired heart disease. In April, 2011, a continuous medical-surgical programme was inaugurated at Clínica Girassol in Luanda. The main goals were to initiate permanent and local delivery of services while training local teams, allowing autonomous medical and surgical management of children with heart disease. *Results:* Between April, 2011 and August, 2015, a total of 1766 procedures were performed on 1682 children. Of them, 1539 had CHD and 143 had acquired heart disease; 94 children underwent interventional treatment. A total of 1672 paediatric surgeries were performed on 1588 children: 1087 (65%) were performed with extracorporeal circulation and 585 (35%) were off-pump. The age distribution of the children was 4.5% (n = 76) neonatal, 40.4% (n = 675) between 30 days and 1 year, and 55.1% (n = 921) over 1 year. There were 76 re-operations (4.5%) due to complications. The 30-day mortality rate was 4.2% (71 patients). Education-wise, several Angolan medical and surgical specialists were trained, allowing near-autonomous cardiac care delivery in children with heart disease. *Conclusion:* An innovative cooperation model between a European and an African centre based on permanent delivery of care and education allowed for effective training of local teams and treatment of children with heart disease in their own environment.

Keywords: CHD; paediatric cardiology department; Sub-Saharan Africa

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In developed countries, advances in paediatric cardiology and surgery have allowed survival into adulthood for the majority of children with congenitally malformed hearts.[1]

The epidemiological patterns of cardiac disease differ enormously between developed countries and Sub-Saharan African nations, where rheumatic heart disease is still a public health problem and CHD has a very bleak prognosis.[2–5] The tremendous advances in diagnostic tools and interventional and surgical solutions for complex CHD have not yet been replicated in Africa, with very few exceptions.[2,6–10]

In the African continent, more than 280,000 newborns/year with CHD are left untreated, validating the natural history of congenital malformed hearts.[1,9,10]

This large number of children with CHD and rheumatic heart disease have very limited or no access to proper medical, interventional, or surgical care, and their future is progression to irreversible cardiopulmonary lesions and death.[6,11]

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Recently, several strategies have been implemented to change the situation by trying to offer and improve medical care in these countries. These strategies include referral to developed countries,[6,12–16] deployment of foreign medical teams on medical/surgical missions[17–19] – so called "medical safaris" – and the development of permanent local teams progressing to achieve minimal dependence followed by full autonomy.[18,20]

Angola has an estimated population of 22 million people, and 46.4% are under 15 years of age.[21] Out of 800,000 births each year, 120,000 children under the age of 5 years die and mortality under 1 year of age is 96 per 1000 live births.[21,22]

The epidemiological profile of these deaths in Angola before the age of 5 is dominated by respiratory infections, acute diarrhoea, and malaria, whereas congenital anomalies are merely the 7th cause in this age group;[23] however, with over 800,000 live births, more than 6500 newborns will have CHD, of which 2000 cases are critical, contributing to a significant cause of mortality and morbidity in the paediatric age.

In Angola, until 2010, interventional/surgical treatment of these patients was always performed abroad, namely in Israel, France, the Republic of South Africa, and Portugal, and never exceeded 50 cases per year.[12]

In April, 2011, a medical-surgical programme aiming to treat CHD and acquired paediatric heart disease was started at Clínica Girassol in Luanda, Angola. This report focusses on reporting our experience in reaching such objectives in a limited human resource environment.

Material and methods

In 2011, a cardiac partnership programme was established between Clínica Girassol owned by SONANGOL – which is a state company in charge of oil resources and natural gas in Angola – the Angolan Ministry of Health, and Hospital da Cruz Vermelha Portuguesa located in Lisboa, Portugal.

The main goal was to develop permanent and local diagnostic clinics for the diagnosis and treatment of children with congenital or acquired heart disease and train local national teams in this area.

The government/Sonangol assumes all financial support for this programme at no cost to patients and their families.

The infrastructure of Clínica Girassol includes a dedicated fully equipped operating room for cardiac surgery, a 12-bed Pediatric ICU, a 16-bed Neonatal ICU, a 20-bed ward, a Cath Lab, a 256-slice CT scan facility, a 3-Tesla MRI facility, an ECHO Lab for 2D, 3D, transoesophageal, foetal, and Doppler procedures, and an Electrophysiology Lab allowing full evaluation and treatment of any kind of pathology except for heart and lung transplant, but including ECMO equipment, for any age group.

The local team has one surgeon, one anaesthetist, one cardiologist, ICU doctors, ward and operating room nurses, one perfusionist, several physiotherapists, and administrative staff.

The Circulation Department of Hospital da Cruz Vermelha has been a national referral centre for the treatment of cardiovascular disease in adults and children since 1998, with particular focus on neonatal procedures. It includes a multidisciplinary team of paediatricians, cardiologists, paediatric cardiologists, cardiac surgeons, intensivists, health technicians, and nurses.

There are members of the Department's staff that have double nationality (Portuguese and Angolan), and others were born and raised in African countries with Portuguese as the official language: namely, Angola, Mozambique, and São Tome e Príncipe.

A permanent Hospital da Cruz Vermelha 13-element team composed of two surgeons, one paediatric cardiologist, one anaesthetist, two scrub nurses, two perfusionists, and five ICU nurses is based in Luanda. A rotational system allowed for the permanent presence of the full multidisciplinary team in Luanda.

The programme was initiated in December, 2010, the first surgery was performed in March, 2011, and the first patient was treated with cardio-pulmonary bypass in May, 2011.

The referral process encompasses pre-evaluation of patients at the Pediatric Hospital in Luanda and/or other paediatric units, followed by admission at Clínica Girassol for detailed investigation and treatment.

This programme underwent some changes with time: first, between April, 2011 and August, 2015, the period we are studying in this paper, the programme began with the complete multidisciplinary Portuguese team present in Angola, and later evolved to a team with one senior Portuguese local consultant surgeon, one paediatric cardiologist, one anaesthetist, one perfusionist, and two ICU nurses based in Luanda.

The risk adjustment for congenital heart surgery (RACHS-1) method was used to adjust for differences in case mix while analysing in-hospital mortality in Angola. To apply this method, cases were assigned to one of six pre-defined risk categories based on the presence or absence of specific International Classification of Diseases, Ninth Revision, Clinical Modification diagnosis and procedure codes; risk category 1 has the lowest risk for death and risk category 6 the highest. Cases with combinations of cardiac surgical procedures – for example, repair of coarctation of the aorta and ventricular septal defect

repair – were placed in the risk category corresponding to the single highest-risk procedure.[24,25]

We have also created a rheumatic heart valve disease outpatient clinic where patients admitted for valve repair or valve replacement are evaluated. The outpatient clinic allows International Normalized Ratio (INR) monitoring by prothrombin time, penicillin administration, and education of both patients and their families about the disease, its implications, and the need for secondary or primary prophylaxis. Warfarin is the anticoagulation treatment used after valve replacement with a monthly follow-up schedule and adherence was seen to be favourable. The distribution of medication involves no cost for our patients.

Patients who live at a distance from the hospital and cannot visit the clinic on a regular monthly basis have a fixed dose adjusted during the hospital stay.

From the beginning, focus was placed on training and integration of Angolan professionals, including cardiac surgeons, cardiologists, anaesthetists, nurses, and technicians, needed for sustainable local team work in the treatment of congenital/acquired heart disease in the paediatric age as well as on collaboration with the departments of physiotherapy, haemotherapy, and pathology. Knowledge was transmitted during full-team medical rounds, weekly discussion of outpatient diagnosis and treatment plans, and while reviewing ECG, chest X-ray, transthoracic or transoesophageal echo, CT angioscan, MRI, and haemodynamic angiography. In addition, a weekly meeting dedicated to a specific disease/theme and morbidity and mortality conferences were organised. Rounds and cardiac team meetings to discuss the best patient management strategy included participation of an Angolan cardiologist, paediatrician, surgeon, microbiologist, nutritionist, and physiotherapist.

Statistical methods

Categorical variables are expressed as frequencies or percentages; continuous variables are expressed as mean ± standard deviation or as medians and ranges, according to distribution. The SPSS20 (IBM SPSS Statistics version 20 × 86 multiple languages) and Origin statistical packages were used.

Ethical standards: All patients gave informed consent for the surgical or interventional procedures and for anonymous treatment of their data for scientific purposes.

Results

Clinical activity

Between April, 2011 and August, 2015, a total of 1766 procedures were performed on 1682 children; there were 1539 children with CHD and 143 with acquired heart disease; 94 children were treated with interventional cardiology and 1588 children were admitted for 1672 surgeries (Fig 1) of these, 1087 (65%) were performed with extracorporeal circulation and 585 (35%) were off-pump.

In 1393 cases (83.3%) corrective surgery was performed, and in 279 cases (16.6%) a palliative procedure was performed as follows: pulmonary artery banding, 132 cases; systemic to pulmonary central shunt, three cases; modified Blalock–Taussig, 115 cases; Glenn shunt, 27 cases; Damus–Kaye–Stansel procedure, two cases.

Female patients were predominant, numbering 869 (52%), versus male patients, who numbered 803 (48%), with a very high incidence of 181 (10.5%) patients having Down's Syndrome (Table 1).

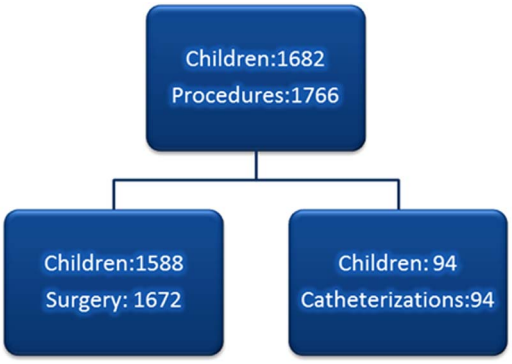

Figure 1.
Total procedures performed on children.

Table 1. Patient characteristics.

Total surgeries	**1672**
CEC	1087 (65)
No CEC	585 (35)
Re-operations:	76 (4.5)
Congenital/acquired	53 (3.7)/23 (16)
Corrective	1393 (83)
Palliative	1279 (17)
Age	
⩽30 days	76 (4.5)
31 days to 1 year	675 (40.4)
⩾1 year	921 (55.1)
Female/male	869/803
Down's syndrome	181 (10.6)
Median days in intensive care [n (range)]	2.4 (1–57)
Congenital/acquired	2.4 (1–54 days)/ 2.2 days (1–9 days)
Median length of stay [n (range)]	9 (1–76)
Congenital/acquired	9 days (2–76 days)/ 12 (5–444)
In-hospital mortality	71 (4.2)
Values are expressed as n (%) unless otherwise indicated	

CEC = corporeal extra circulation

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The age distribution was as follows: 76 (4.5%) neonatal (700 g to 4.4 kg, mean 3.4 kg), 675 (40.4%) between 30 days and 1 year (2.0–10 kg, mean 4.9 kg), and 921 (55.1%) over 1 year of age (4.5–56 kg, mean 15.6 kg) (Fig 2).

In terms of age distribution, with time we documented a trend for more children being diagnosed at an increasingly younger age every year (Fig 3).

There were 1539 cases with congenital cardiac disease, and the group with a left-to-right shunt was predominant (n = 1020), followed by the group with right (n = 316) and left heart lesions (n = 252). The most frequent conditions in each of these groups were ventricular septal defects, tetralogy of Fallot, and coarctation of the aorta (Figs 4 and 5).

A total of 143 children had acquired heart disease, mostly rheumatic valve disease. Among children with valve disease, 92 had mitral insufficiency, 35 had mitral regurgitation and stenosis, and one had mitral stenosis. In all, 15 children had aortic mitral disease, 58 had undergone valve repair, 70 had undergone valve replacement with mechanical prosthesis, and 15 had undergone double valve replacement - of both aortic and mitral valves. Of these, 38 patients were also subjected to tricuspid repair, mostly using the De Vega technique.

The choice between valve repair and valve replacement was dependent on leaflet pliability and the extent of sub-valvular involvement in the rheumatic disease process, with valve repair being the preferred method.

Most procedures were performed in the RACHS-1 risk categories 2 and 3, representing 76% (n = 1329) of the total number of surgeries (Fig 6).

The median duration of ICU stay was 2.4 days for CHD, with a range of 1–57 days, and 2.2 days for acquired heart disease, with a range of 1–9 days; the median length of hospital stay was 9 days, with a range of 2–76 days, and 12 days, with a range of 5–44 days, respectively, in these two groups.

There were 200 cases of postoperative complications in 1539 congenital procedures (12.9%) and 40 cases in acquired heart diseases (27.9%).

Re-operation during the same hospital admission was needed in 23 cases of acquired heart lesions (16%) due to mitral valve repair failure (n = 21), aortic valve repair failure (n = 1), and prosthetic mitral valve thrombosis and a thromboembolic event (n = 1). In the congenital group, re-operation was needed in 53 cases (3.4%) because of residual shunts (n = 17), Blalock–Taussig shunt occlusion (n = 10), residual gradient (n = 7), haemorrhage (n = 3), chylothorax (n = 2), and other causes (n = 13). Other complications for congenital heart lesions included pericardial/pleural effusion (n = 69, 4.4%), nosocomial infections (n = 58, 3.7%), postoperative arrhythmia – which occurred in 29 cases (1.9%), in which two cases required a permanent pacemaker

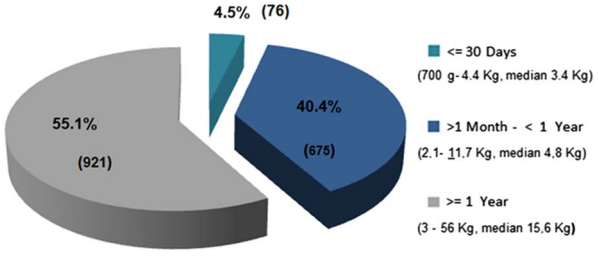

Figure 2.
Age and weight distribution of patients.

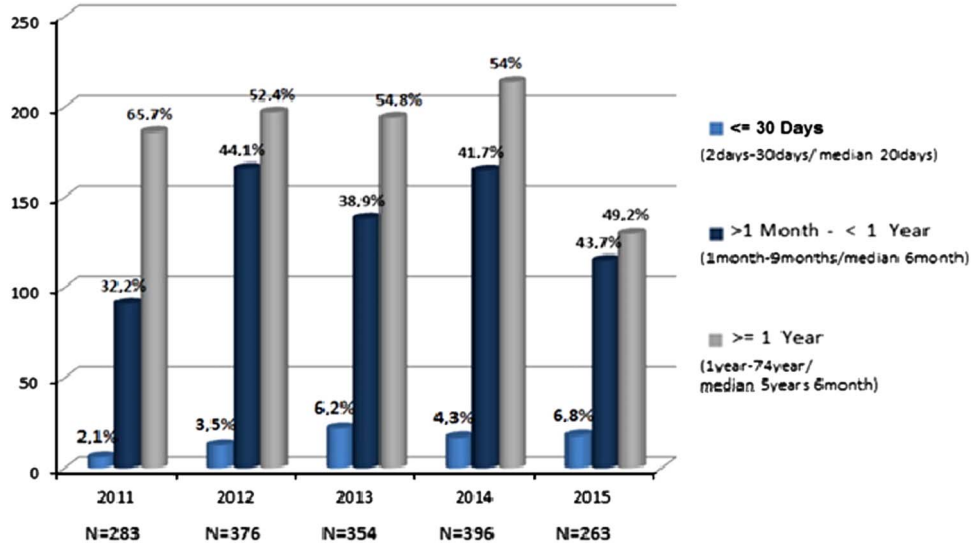

Figure 3.
Distribuition of the various age groups by year (April, 2011 to August, 2015).

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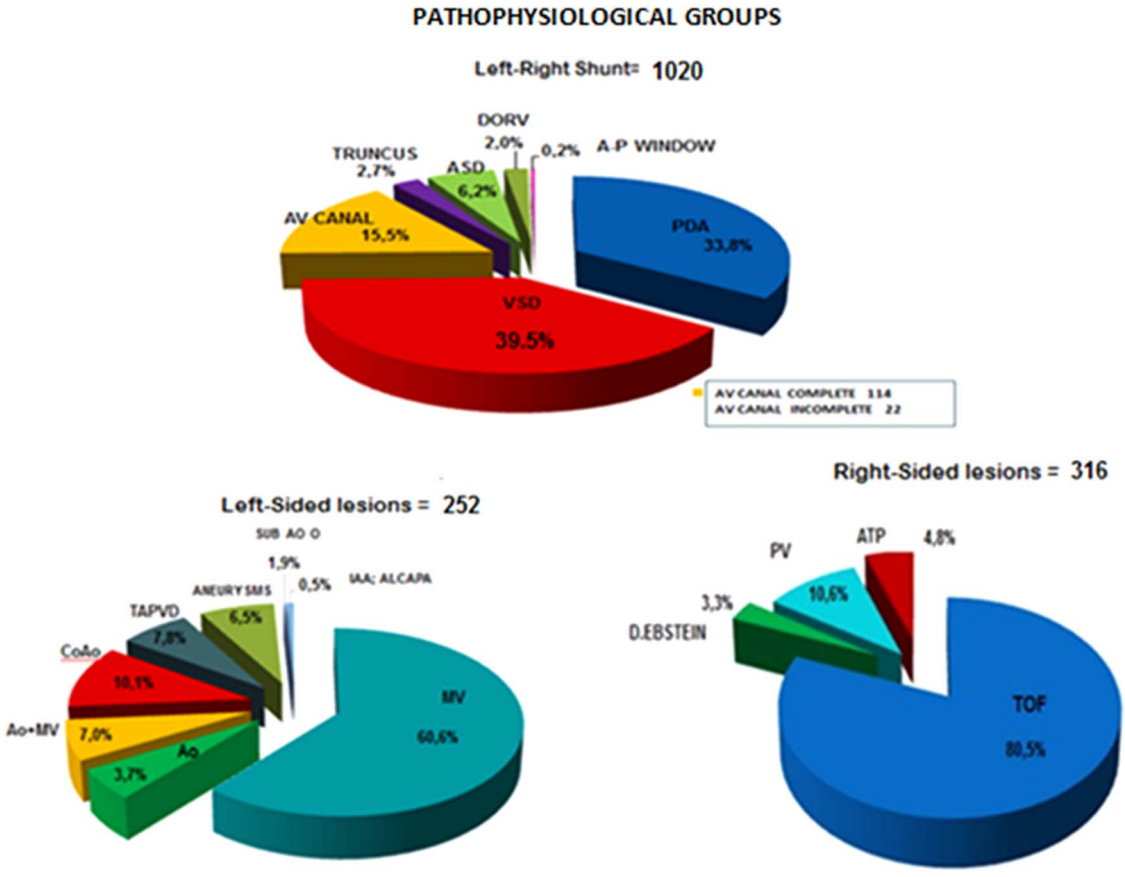

Figure 4.
Distribution of heart diseases by pathophysiological groups. ALCAPA, anomalous left coronary artery from the pulmonary artery; Ao, aortic valve; A-P window, aortopulmonary; ASD, atrial septal defect; ATP, pulmonary atresia; AV canal, atrioventricular canal; DORV, double outlet right ventricle; IAA, interrupted aortic arch; MV, mitral valve; PDA, patent ductus arteriosus; PV, pulmonary valve; Sub-Ao-O, obstacle sub-aortic, aneurysm, atrial septostomy; TAPVD, total anomalous pulmonary venous drainage; TGA, transposition of great arteries; VSD, ventricular septal defect.

for postoperative AV block – and haemorrhage (n = 20, 1.3%). For acquired heart lesions the complications were pericardial/pleural effusion (n = 5, 3.49%), nosocomial infections (n = 2, 1.4%), postoperative arrhythmia – which occurred in 20 cases, in which one required a permanent pacemaker for postoperative AV block – and haemorrhage (n = 7, 4.9%).

Total in-hospital mortality was 0.7% (one patient) in the acquired disease group due to left ventricular dysfunction and 4.5% (70 patients) in the congenital disease group due to sepsis (n = 15), low cardiac output (n = 8), multiple organ dysfunction (n = 10), respiratory failure (n = 14), sudden death (n = 9), haemorrhage (n = 9), and pulmonary hypertension (n = 5). Mortality was higher in RACSH-1 groups 2 and 3. (Fig 6).

A total of 94 children, of whom 67 were female and 27 were male, were treated in the Cath Lab by percutaneous intervention, with no mortality or morbidity (Table 2). The median age of these patients was 4.7 years and ranged from 3 days to 19 years.

Training

We began our training programme by enlisting a locally trained military surgeon in 2011 for further training in paediatric cardiac surgery. He had previous training in general surgery and short-term cardiothoracic surgery. In 2014, he became the Head of General and Cardiothoracic Surgery at the Hospital Militar de Luanda and maintains locum activity at Clinica Girassol. Our next two trainees joined the programme in 2014, one from Angola and one from Portugal. Both are cardiothoracic surgeons seeking further training in paediatric cardiac surgery and they are close to completing a 2-year and a 6-month programme, respectively, in paediatric cardiac surgery. They became fully fledged paediatric cardiac surgeons with the Clínica Girassol team. The Angolan trainee performed 243 major surgeries from August, 2014 to February, 2017 as a first surgeon. The Portuguese trainee performed 60 surgeries in 6 months. They are able to perform all corrective heart surgeries beyond the neonatal period independently.

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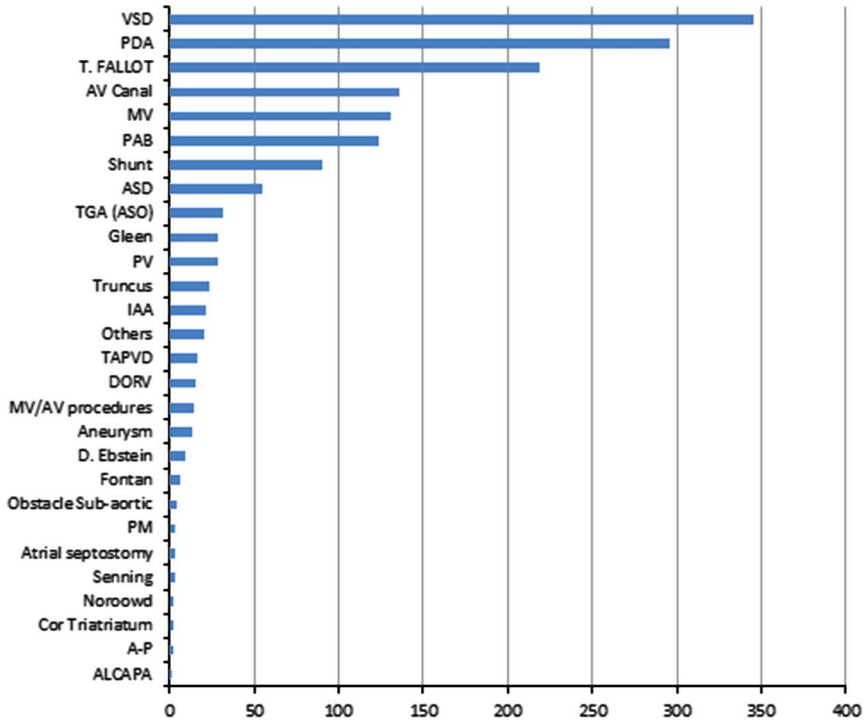
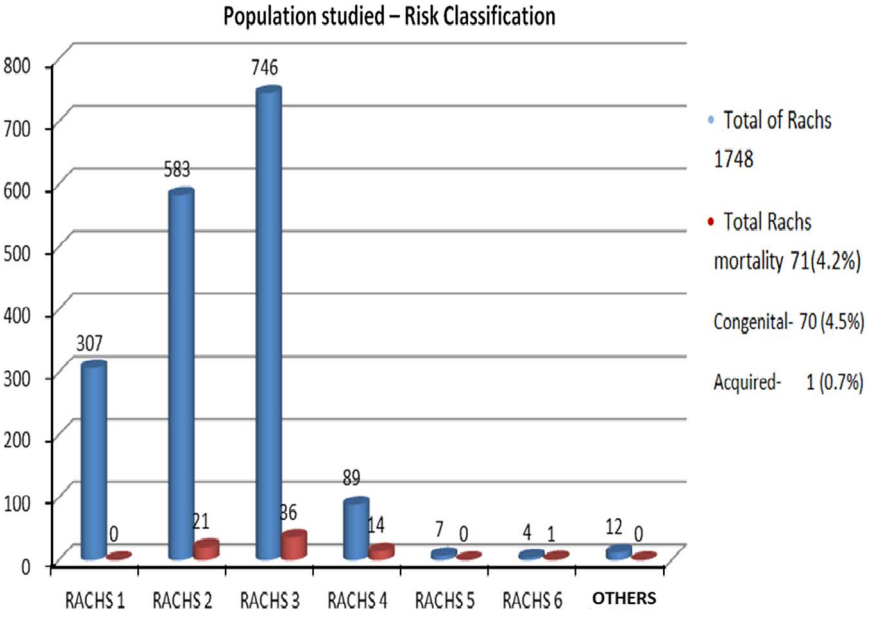


Figure 5.
Specific surgical procedures performed. ALCAPA, anomalous left coronary artery from the pulmonary artery; A-P window, aortopulmonary; ASD, atrial septal defect; ASO, atrial switch operation; AV canal, atrioventricular canal; DORV, double outlet right ventricle; IAA, interrupted aortic arch; MV, mitral valve; MV/AV procedures, aortic valve procedures; PAB, pulmonary artery band; PDA, patent ductus arteriosus; PM, pacemakers; PV, pulmonary valve; Sub-Ao-O, obstacle sub-aortic, aneurysm, atrial septostomy; TAPVD, total anomalous pulmonary venous drainage; TGA, transposition of great arteries; VSD, ventricular septal defect.



Figuer 6.
Risk classification and mortality using RASH-1.

Our programme also included the training of an anaesthesiologist, a perfusionist, and two paediatric cardiologists.

The anaesthesiologist, already experienced in adult cardiothoracic surgery, became competent enough to perform all interventions on her own in 1 year. She

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Table 2. Interventional procedures performed.

Catheter interventional treatment (n = 94)	
Closure of PDA	41
Valvular pulmonar stenosis	27
Closure of ASD	3
Atrioseptostomy of rashkind	14
Closure of MAPCA	1
Pulmonary artery branch stenosis	3
Obstruction of shunt	2
Valvular aortic stenosis	2
Coartaction of aorta	1

ASD = atrial septal defect; MAPCA = major aorto-pulmonary collateral arteries; PDA = patent ductus arteriosus

performed 100 anaesthetic procedures for cardiac surgery, including on neonates, completely independently during the period of this study.

In 2013, the paediatric cardiologists initiated their training in Pediatrics for a 2-year period and then moved to a 3-year training programme in Pediatric Cardiology, thus allowing them to acquire the requisite expertise in the care of children with heart disease in multidisciplinary teams.

We have also trained one experienced adult haemodynamic cardiologist and two haemodynamic technicians adequately to be able to perform the most common interventions without supervision of the Portuguese consultants after the initial 1 year.

We are also developing Pediatric Cardiology and Cardiac Surgery Residency Programs.

Discussion

Since the beginning of the programme in April, 2011, until August, 2015, 1682 children have been treated at Clínica Girassol. All 800 children in the previous waiting list were treated during the first 2 years of the programme. Hospital mortality was 4.2%, with 0.7% in acquired and 4.5% in CHDs.

For children with operable CHD, early primary repair is preferred over palliation whenever possible. There are a number of aspects to be taken into account when making this decision as they impact not only the treatment but also the results. Socio-economic background, the spectrum of disease, and management issues largely differ from the European pattern.

Late presentation or late diagnosis becomes critical when it comes to malformations that require early correction, such as transpositions, ductus-dependent conditions, and left or right obstacles. Many of these children die before diagnosis or are inoperable at presentation. This might be one of the reasons why only 4.5% of our patient population – namely, 76 patients - was aged <28 days.

Results are equally influenced by a number of comorbidities such as nutritional deficiencies, low birth weight, preoperatory infection, and HIV infection. Although Clínica Girassol is structured and fully equipped with the latest technological advances, limited human resources and a precarious socio-cultural background hamper ideal treatment and adherence to the postoperative care needed for children with complex heart disease.

Rheumatic heart disease is still a calamity in the African continent, and the majority of patients present late in their disease course.[2] Valve repair is always favoured when feasible; in our series of patients with mitral disease, 58 were subjected to valve repair, 70 to a prosthetic heart valve, and 15 to a double mitral and aortic prosthetic valve. Heart valve replacement is reserved for patients in whom repairs fail or are not possible. Warfarin is the anticoagulation treatment employed after valve replacement, with a monthly follow-up schedule, and adherence is favourable. The distribution of medication involves no cost for our patients.

Patients who live at a distance from the hospital and cannot visit the clinic on a regular monthly basis have a fixed dose adjusted during the hospital stay. Our experience with prosthetic valve thrombosis and thrombotic events after mechanical valve replacement in this study cohort is limited to one case, and increased INR due to misunderstanding/mistakes in anticoagulation medication was documented in several cases in our series. None of these patients suffered serious complications.

We believe that the prevalence of rheumatic heart disease in Angola is identical to that in the rest of Africa, which is unacceptable. All effort should be made to guarantee that preventive measures are applied, and this should be undertaken by the government and by all interested parties.

In this context, we have created a rheumatic fever outpatient consultation where we promote primary and secondary prevention, which is fundamental for eradication of rheumatic heart disease.

So far, in Africa, only South Africa has national guidelines for the prevention of rheumatic heart disease.[2,26,27]

The initiation of a national programme that contemplates primary and secondary prevention of rheumatic heart disease is mandatory. Its integration in specific African programmes is also pivotal.

A total of 94 patients were treated by interventional heart catheterization with no morbidity or mortality. Initially, these interventional procedures were performed with assistance from a local interventional adult cardiologist trained in Portugal. As the local team gained experience, the Portuguese teams assisted the local team in performing the

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interventions. In later years, the local team was able to perform simple interventions independently, with the team from Hospital da Cruz Vermelha being available for any support or advice if needed. Eventually, one cardiologist, three nurses, and two haemodynamic technicians were trained to perform the most common interventions without supervision. Supervision was aimed at more complex procedures. Currently, we have reduced the number of nurses and haemodynamic technicians, transferring full responsibility to the local team in the majority of situations. Training an independent interventional paediatric cardiology team and implementing an interventional paediatric cardiology service in a Sub-Saharan setting is challenging but achievable.[15] The low number of interventional heart catheterization procedures compared with surgery is due to the fact that many of the referred patients have considerable left–right shunts that preferably require surgery; however, with improved diagnostic acuity and experience we strongly believe that this number will increase in the future.

Very few African countries have the resources to provide optimum paediatric cardiac care, and large numbers of families with CHD lack the resources to access the available treatment.

Adequate finance is pivotal and local personnel must be trained.

This Angolan cardiothoracic health care programme for children involved a heavy government commitment towards infrastructural requirements and financing for those in need - that is, the poorest patients. These aspects are desperately lacking in the majority of Sub-Saharan Africa.[28] As a result, Angola has been successful in saving neonates with CHD who would have otherwise died waiting for funding in other African countries with cardiac programmes. This is due to financial constraints, which resulted in the death of 32.5% of patients on a waiting list in Cameroon.[29]

Unit "twinning" has been proposed rather than the relatively inefficient medical safaris. Our model, as R. H. Kinsley called it, is a real "marriage", and we must bear in mind that the matrimony should be appropriate - for example, Angola/Mozambique and Portugal, and French-speaking European countries and West Africa.[1]

True Heart Team integration, fostering multidisciplinary collaboration, and intense exposure to congenital and acquired heart disease allowed successful training of multiple Angolan specialists, including cardiac surgeons, paediatric cardiologists, anaesthetists, intensivists, nurses, and cardiopneumologists, who can now, under supervision, independently perform surgical and interventional procedures in children with heart disease.

Conclusion

A unique cooperation model between a European and an African centre from Portugal and Angola has successfully allowed the treatment of numerous children with heart disease in their own environment as well as the training of local teams with the advantage of common language and common history.

The success of this programme clearly demonstrates the benefit of treating African children with heart disease in their local environment and highlights the importance of the contribution of the government and all interested parties towards a favourable outcome.

Looking into the future, the challenge is the creation of a national referral network in order to structure and amplify cardiac assistance to every child included in the Angolan health system all over the country.

We have made enormous progress in the three areas that constitute our core mission: service, training, and research.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation set by the Health and Disability Ethics Committees and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the Cruz Vermelha Health Board Research Review Committee.

Supplementary materials

For supplementary material referred to in this article, please visit https://doi.org/10.1017/S1047951117001202

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