

## EXPERIENCE WITH PALLIATIVE PROCEDURES FOR CONGENITAL CYANOTIC HEART DISEASES IN GHANA: A 20-YEAR REVIEW

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### Abstract

**Background:** Currently, in the management of congenital cyanotic heart diseases with favourable anatomy, primary correction is the preferred choice. But palliative procedures have still maintained an important role in resource-poor regions of the world. We sought to analyze our institutional results for such palliation over a 20-year period. The purpose of the study was to provide information that would guide both practitioners and referring doctors on the future management of congenital cyanotic heart diseases, and also to provide data for comparative studies in the sub region in the future.

**Patients and Methods:** A retrospective study was carried out in the National Cardiothoracic Centre involving all patients who had palliative procedures for congenital cyanotic heart diseases from January 1992 to December 2011.

**Results:** There were 264 patients, 59% males and 41% females. The case load increased from 2 cases per year in the first year to 34 in the last year of the study. The age range was 4 months – 42 years, with a mean of  $7.1 \pm 6.9$  years. Tetralogy of Fallot comprised 257 (97.4%) of the cases, tricuspid atresia 4 (1.5%), and double outlet right ventricle 3 (1.1%). The modified Blalock-Taussig shunt (MBTS) was performed in 262 (92.2%), and the Waterston shunt in 2 (0.8%) of the cases. The overall complication rate was 11.8%, comprising shunt occlusion 7.6%, bleeding requiring re-exploration 0.8% and early mortality of 3.4%.

**Conclusion:** The modified Blalock-Taussig shunt provided good palliation for congenital cyanotic heart diseases in this environment. We consider it a suitable management alternative when financial and logistic constraints delay primary repair in resource-poor settings.

**Key Words:** Congenital cyanotic heart diseases, Palliative procedures, Good outcome, Ghana

### Introduction

Children with congenital cyanotic heart diseases faced a very bleak future until Alfred Blalock performed the first subclavian artery-to-pulmonary artery anastomosis in 1944 on a severely cyanosed 15-month old girl<sup>1</sup>. The procedure, in its classical form, involved anastomosis of the divided subclavian artery to the ipsilateral branch pulmonary artery in end-to-side fashion. The Blalock-Taussig shunt (BTS), as it became known, immediately became popular due to its several advantages. Subsequently, many other palliative shunts were developed by other investigators.

The Pott's shunt involves an anastomosis between the descending thoracic aorta and the left pulmonary artery<sup>1</sup>. The Waterston shunt involves an anastomosis between the ascending aorta and the right pulmonary artery<sup>1</sup>. The Davidson shunt involves an anastomosis between the ascending aorta and the main pulmonary artery<sup>1</sup>. And the Glenn shunt involves an anastomosis between the superior vena cava and the right

pulmonary artery<sup>1</sup>. The BTS was subsequently modified by de Leval in 1981<sup>2</sup>, by interposing a polytetrafluoroethylene (PTFE) graft between the subclavian artery and the ipsilateral pulmonary artery. This avoided division of the subclavian artery and its attendant complications. The modified Blalock-Taussig shunt (MBTS) of de Leval, has since been the most frequently performed palliative procedure for congenital cyanotic heart disease<sup>1</sup>.

The current trend in the management of cyanotic congenital heart disease with a favorable anatomy is early primary repair. However, in resource-poor regions of the world, this strategy is not always feasible due to logistic and funding constraints. Palliative procedures have thus maintained their role in the management of such patients. The rationale for a palliative shunt is to increase pulmonary blood flow for improved oxygenation. The result is amelioration of cyanosis and its associated complications. Accompanying such improvement is a significant increase in effort tolerance, and over time, growth in size of the pulmonary vasculature occurs. The changes following palliation are advantageous for the outcome of definitive repair.

The aim of this study is to analyze the outcome of palliation for congenital cyanotic heart diseases in our institution over a 20-year period. The importance of this study is that it will guide us and other institutions in the sub region in the subsequent management of

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Conflict of Interest: None declared

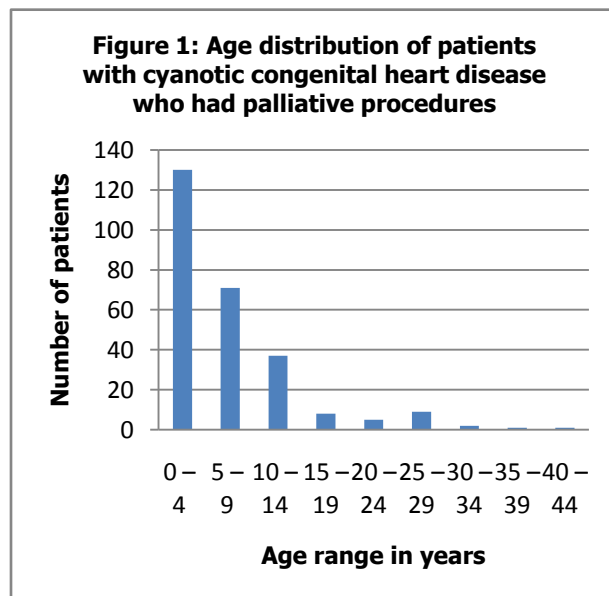
patients with congenital cyanotic heart diseases. It will also provide data for comparative studies in the future.

## Patients and Methods

All patients who had palliative procedures for cyanotic congenital heart disease in the National Cardiothoracic Centre from January 1992 to December 2011 were entered into a retrospective study. The data were obtained from our institutional records database and the patients' case notes. The data included the history, physical examination, investigations, treatments and complications. The analysis for means, frequencies, and standard deviations was performed using Microsoft excel 2010 statistics software, Windows 7.

## Results

There were 264 patients, with 156 (59%) of them being males while 108 (41%) were females, with a male to female ratio of 1.4: 1. The case load increased from 2 cases in the first year to 34 in the last year of the study. The age range was 4 months – 42 years, with a mean of  $7.1 \pm 6.9$  years, and median of 5 years. The majority 130 (49.2%) of the patients were 0 – 4 years old. The age distribution is shown in Fig. 1



**Fig. 1:** Age distribution of the patients

In all, 257 (97.4%) of the cases were diagnosed with tetralogy of Fallot. The rest were tricuspid atresia in 4 (1.5%) patients and double-outlet right ventricle in 3 (1.1%). Two hundred and sixty-two (99.2%) MBTS were performed as against 2 (0.8%) Waterston shunts. Twenty-two (8.4%) of the 262 MBTS were performed on the left side; the rest were right-sided. The overall complication rate was 11.8%, comprising shunt occlusion in 7.6%, bleeding requiring re-exploration in 0.8% and early mortality of 3.4%. The constructed

shunts remained patent for  $2.3 \pm 1.9$  years (1 week – 5.5 years, median 1.8 years).

## Discussion

Two patients underwent palliation in the first year of the study, and 34 in the last year. This significant increment was due to the increase in the population of the country and referrals from other countries in West Africa. But more so, from increased referrals due to greater caretaker awareness, increased funding sources, better health education among the population, and other factors. The male to female ratio was 1.4: 1. The youngest patient was 4 months old and the oldest was 42 years. Both of them had tetralogy of Fallot. The modal age group was 0 – 4 years. This was when the disease conditions were most symptomatic, and also when the parents could afford the cost of the procedure. Majority (97%) of the cases were tetralogy of Fallot, the reason being that tetralogy of Fallot is the commonest cyanotic congenital heart disease<sup>3,4</sup>. Almost all the shunts (99.2%) were Modified Blalock-Taussig shunts. This is because of the advantages of the Blalock-Taussig shunt over the others, notably being the suitability of the blood flow through the shunt, which is determined by the subclavian artery blood flow and the less tendency to develop congestive cardiac failure, as opposed to the Potts and Waterston shunts. These and other features made the BTS the most effective palliative procedure<sup>5</sup>. Even a 60-year survival of the BTS had been reported.<sup>6</sup> Only two Waterston shunts were performed. These were during the earlier part of the study. The shift towards the BTS is in accordance with its better outcome<sup>5</sup>.

The MBTS was offered to the patients with frequent hypercyanotic attacks, high haematocrits and narrow branch pulmonary arteries. The procedure was performed through a thoracotomy, entering the chest through the fourth intercostal space, retracting the lung inferiorly, and dissecting out the subclavian artery and the branch pulmonary artery. A PTFE graft about the size of the subclavian artery was chosen and interposed between the subclavian and branch pulmonary arteries. Most of the MBTS (91.6%) were performed on the right. This is due to the relative ease of dissection and takedown of the MBTS on the side contralateral to the aortic arch at the subsequent intra-cardiac repair. When preoperatively, a right sided aortic arch was diagnosed, the shunt was performed on the left. Re-do shunts were also performed on the left.

The overall complication rate was 11.8%, comprising shunt occlusion in 7.6%, early mortality in 3.4% and bleeding requiring re-exploration in 0.8%. The mean duration of the shunt occlusion was  $2.3 \pm 1.9$  years. The causes of shunt occlusion are usually considered as early (within 30 days) or late (after 30 days). The early occlusion can be due to patient factors, graft factors or surgical technique. An example of a patient factor is a high haematocrit leading to a high viscosity which then leads to early shunt thrombosis.

This problem is solved by venesection intra-operatively. Graft factors can be due to a soft graft that collapses easily, a long graft that kinks, or a relatively small graft that thrombosis easily. Good surgical technique includes choosing the appropriate vessel, and also careful construction of the anastomosis. The late occlusion factor is a patient factor, which is due to the child outgrowing the shunt and thereby rendering the flow relatively insufficient, resulting in a gradual increase in the haematocrit, and finally leading to shunt thrombosis. The mean occlusion duration of 2.3 years reinforces the opinion that total intra-cardiac repair after an MBTS should be less than 2 years after the shunt (that is, before the shunt gets occluded). Re-do MBTS were done for the blocked shunts. Perigraft seroma has not been observed in this study, though some other studies have reported the complication<sup>7,8</sup>. The early mortality of 3.4% is comparable to that of Singh et al, of 4.5%<sup>9</sup>. The causes of our mortality were desaturation and deterioration after the procedure in 1.9%, clotted haemothorax in 0.8% and pleural effusion after discharge from hospital in 0.8%. The long term complications of the MBTS could not be assessed reliably in this study because of the limitations of the study.

**Limitations of the study:** The medium and long term complications of the study could not be estimated reliably because some of the patients' case notes could not be assessed. This problem is currently being solved by improved record keeping, and also by computerizing the data in the case notes.

## Conclusion

The modified Blalock-Taussig shunt provided good palliation for cyanotic congenital heart disease. We consider it a suitable management alternative when financial and logistic constraints delay primary repair in resource-poor settings.

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