A Retrospective Study of the Clinical Outcomes Following Surgical Correction of Supra Cardiac Total Anomalous Pulmonary Venous Connection

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Abstract: Total anomalous pulmonary venous connection is a rare (1% of children born with congenital heart diseases), life threatening congenital cardiac anomaly. In this study we review and discuss the presentation, clinical features, investigations and factors, which influence the surgical outcome in patients with total anomalous pulmonary venous connection who underwent surgery at Madras Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019.

Keywords: Total anomalous, pulmonary, congenital

1. Introduction

Total anomalous pulmonary venous connection is a rare (1% of children born with congenital heart diseases), life threatening congenital cardiac anomaly. It was first described by Wilson (1) in 1798. Friedlowsky in 1886 described it as a pathologic entity. But it was not until 1950 (2) that the first case was reported in which the clinical diagnosis was made, and this was accomplished through the use of cardiac catheterization. Muller (3) in 1951 was the first to repair it successfully. Since then there has been progressive improvement in operative results. However, the surgical mortality varies from 8% to 20% in most series (4, 5, 6). The mortality rate is higher in infants. The causes for the higher surgical mortality in this group may include pulmonary congestion, elevated pulmonary vascular resistance, and the critical condition of many infants at the time of operation. Thus early accurate diagnosis, prompt surgical intervention and careful preoperative and postoperative care are the key to success.

In this study we review and discuss the presentation, clinical features, investigations and factors, which influence the surgical outcome in patients with total anomalous pulmonary venous connection.

2. Aims and Objectives

To retrospectively evaluate presentation, clinical features, investigations and factors which influence the surgical outcome in patients with total anomalous pulmonary venous connection who underwent surgery at Madras Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019.

3. Methods and Materials

Patient Population
22 patients who underwent surgery for supracardiac total anomalous pulmonary venous connection at Madras Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019.

Post Operative Management
This included prolonged elective ventilation. We used only pressure control mode of ventilation. We maintained partial pressure of carbon dioxide below 30mmHg. The duration of ventilation ranged between 1 and 8 days with a mean of 2.18 days.
Weaning Protocol
All patients were electively ventilated for a minimum of 24 hours. If patients were haemodynamically stable weaning was started. Frusemide was given approximately 3 hours before extubation. They also received one dose of steroids prior to extubation.

Mean ICU stay was 3.45 days. The mean ICU stay among those who survived was 3.54 days. Survivors were discharged from the hospital after a mean interval of 9.23 days (range, 6 to 15 days).

Echocardiographic and Cardiac Catheterisation Studies
Echocardiography was done for 18 patients while cardiac catheterization was done for 6 patients. Most of our diagnosis was echo based.

4. Data Analysis

The SPSS version 11 statistical program was used to analyze results. Chi square test was used for discrete variables.

5. Results

The operative mortality rate for isolated TAPVC was 40.9% (9 of 22 patients).

The major cause of death was pulmonary hypertensive crisis 67% (6 of 9 patients). The other causes of death were low cardiac output, congestive cardiac failure and electrolyte imbalance.

Among the 22 patients, 13 patients weighed less than 10kgs. Of the 9 patients who died, 8 weighed less than 10kg, which was statistically significant (p=0.021).

Risk factors for early death were analyzed by univariate analysis. Age less than 12 months, weight less than 10kg, size of ASD less than 10mm, and preoperative failure were significant risk factors for postoperative mortality. Of the 22 patients, 13 patients were below the age of 1 year and 9 were above 1 year. Also there were 8 patients who were below the age of 12 months among the 9 patients who died. This was statistically significant (p=0.009).
From the graph depicting the 95% CI for BSA, those who died had a significantly lower BSA. The BSA for those who died ranged from 0.21 to 0.72 (mean 0.35), while for those who were alive it ranged from 0.22 to 1.65 (mean 0.84).

Sixteen patients were diagnosed to have clinical cyanosis. However there was an equal distribution between the two groups.

<table>
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<th>Total</th>
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p=0.157

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p=0.245

Nine patients were in CCF. Of these five patients died. The size of the liver was an estimate of the degree of the heart failure. The larger the size of the liver, the greater was the mortality. Statistically this was not significant. This could be attributed to the small sample size. However the trend was evident.

<table>
<thead>
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p=0.548

The size of the atrial septal defect had a significant bearing on the surgical outcome. Of the 9 patients who died, 7 had an ASD, which measured less than 10mm (p=0.004). The mean of the ASD among the mortality group was 5.21mm while that in the other group 12.23mm.

<table>
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<td>7</td>
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<tr>
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p=0.004

In the group, which underwent circulatory arrest, the total pump time ranged between 42min and 134min (mean 95.62min). The total cross clamp time ranged between 23min and 79min (mean 54.31min).

In the group, which was operated on with moderate hypothermia, the total pump time ranged between 60min and 156min (mean 100.67min). The total cross clamp time ranged between 41min and 53min (mean 49.67min).

All patients required postoperative inotropic support. The mean duration of ventilation was 2.18 days (range 1 to 8 days).

Follow-up ranged from 6 to 60 months. Postoperative follow up was not possible in 3 patients. Surviving patients had normal growth and were free of symptoms

6. Conclusion

Surgery for TAPVC still carries a high mortality.
The conditions contributing to this are: 1. weight at surgery less than 10kgs 2. age at surgery less than 12 months 3. size of ASD less than 10mm 4. late recognition and referral for surgery. It is important therefore to identify patients with TAPVC early and subject them to a corrective procedure at the earliest.

References