Outcome after surgery for pulmonary atresia with ventricular septal defect, a long-term follow-up study

Erik Wiezell1 | Janus F. Gudnason2 | Mats Synnergren2 | Jan Sunnegårdh2,3

1Department of Paediatrics, Södra Älvsborg Hospital, Borås, Sweden
2Children's Heart Center, The Queen Silvia Children's Hospital, Sahlgrenska University Hospital, Gothenburg, Sweden
3Institute of Clinical Sciences, Gothenburg University, Gothenburg, Sweden

Correspondence
Erik Wiezell, Lindebergsgatan 6, SE-506 36 Boras, Sweden.
Email: erik.wiezell@gmail.com

Abstract
Aim: To study the long-term outcome after surgery for pulmonary atresia and ventricular septal defect (PA-VSD), and to determine association between the contribution of major aorto-pulmonary collateral arteries (MAPCAs) to the pulmonary blood flow, comorbidity and cause of death.

Methods: Patients who had undergone surgery for PA-VSD from January 1st 1994 to December 31st 2017 were studied retrospectively. Survival was cross-checked against the Swedish National Population Register.

Results: Seventy patients were identified, giving an incidence of 5.3 newborns per 100 000 live births. In 41 patients (59%) the pulmonary blood flow originated from a patent ductus arteriosus (PDA), while 29 patients (41%) had contribution of the pulmonary blood flow from MAPCAs. Extracardiac disease was found in 34 patients (49%), 16 of whom had 22q11-microdeletion syndrome (23%). Survival at follow-up was similar in patients with and without MAPCAs (72.4% vs. 75.6%, n.s.), with a median follow-up time of 14.3 years (3.2–41.8 years). No difference was found in mortality in patients with or without any syndrome or extracardiac disease.

Conclusion: Long-term survival did not differ between those with and without MAPCAs and no difference in mortality was seen in patients with and without concomitant extracardiac disease or any kind of syndrome.

KEYWORDS
22q11-microdeletion syndrome, congenital heart defect, major aorto-pulmonary collateral arteries, pulmonary atresia with ventricular septal defect, pulmonary blood flow

Introduction
Pulmonary atresia with ventricular septal defect (PA-VSD) with or without major aorto-pulmonary collateral arteries (MAPCAs) is an uncommon congenital malformation, with an incidence of 4–10 per 100 000 liveborn children, accounting for about 2.5% of congenital cardiac defects.1–3 Genetic and environmental factors seem to play a role in the development of this congenital heart defect, but the precise mechanism is still unclear.4 The association with the 22q11-microdeletion syndrome is well-known, and is reported in up to 41% of all the PA-VSD patients, and in up to 65% of patients with MAPCAs.5–7 In patients with 22q11-microdeletion syndrome 75% have a congenital heart defect, and of these about 24% have PA-VSD with or without MAPCAs.8

Abbreviations: PA-VSD, Pulmonary atresia and ventricular septal defect; MAPCAs, Major aorto-pulmonary collateral arteries; PDA, Patent ductus arteriosus.

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Patients with PA-VSD can be diagnosed prenatally by ultrasound, but it can be difficult to map the pulmonary blood supply.\(^9\) The pulmonary vascular supply in patients with MAPCAs varies regarding origin, number, size, course and the part of the lung supplied, and is thus demanding for all the health care professionals involved.\(^10\) Cardiac catheterisation and/or multi row detector computed tomography can be used once the child is born to visualise the anatomy before surgery.\(^11\) The optimal treatment for this malformation is biventricular repair with closure of the ventricular septal defect to create blood flow from the right ventricle to the pulmonary arteries through a conduit with an acceptable right ventricular pressure. In order to repair the defect, initial palliative strategies, such as inserting an aorto-pulmonary shunt or a right ventricle to pulmonary artery connection, are required.\(^12\) Survival without surgery is limited to a few years in the majority of patients PA-VSD, with or without MAPCAs.\(^13\) Several studies have been presented on postoperative results and follow-up of PA-VSD patients.\(^2,3,13\) In recent years, extensive research has been performed to determine how the most favourable results can be achieved in patients with PA-VSD and contribution to the pulmonary blood flow from MAPCAs.\(^3,10,14,15\) We here report the results on long-term survival, associated comorbidities and causes of death in patients with PA-VSD with or without MAPCAs in our hospital.

2 | SUBJECTS AND METHODS

2.1 | Study population

All patients who had undergone surgery due to PA-VSD from January 1st 1994 to December 31st 2017 at the Queen Silvia Children’s Hospital, Sahlgrenska University Hospital were included. Since 1994, paediatric cardiac surgery has been performed at two centres in Sweden, Lund University Hospital and the Queen Silvia Children’s Hospital in Gothenburg\(^16;\) there is no private alternative for this treatment. Each centre has approximately 5 million inhabitants in its referral area. The study was performed as a retrospective study, using the Queen Silvia Children’s Hospital paediatric cardiothoracic database and patient’s medical records at local hospitals.

2.2 | Data collection

Patients with PA-VSD were identified from the paediatric cardiothoracic database at the hospital, and important clinical data were retrieved by manually searching the patient’s original medical records. The diagnoses, including the source of pulmonary blood flow supply, were confirmed in all patients through catheterisation, angiography and surgical reports, as well as data from echocardiography and computed tomography examinations. Survival was cross-checked against the Swedish National Population Register as of 1st May 2020, providing reliable and complete data. All medical records were retrieved with no patient lost to follow-up. The study was approved by the Regional Ethics Committee (No. 518-16, T1123-17).

2.3 | Statistical analysis

The primary outcome was all-cause mortality; secondary outcomes were extracardiac diseases, including syndromes, reoperation and catheter interventions. Differences between groups were studied using the chi-squared test and Student’s t-test. Survival was estimated using Kaplan–Meier curves and the log-rank test to compare survival between groups using IBM SPSS Statistics, version 23. A p-value ≤0.05 was considered statistically significant.

3 | RESULTS

Between January 1st 1994 and December 31st 2017, 70 patients with PA-VSD had undergone surgical treatment at our hospital. In 12 cases, children born before 1994 had been given surgical treatment before 1994, while 58 patients were born during the study period. According to the Swedish National Board of Health, the total number of live births in the referral area covered by the hospital during the study period was 1 091 406 (Statistics Sweden, www.scb.se), giving an incidence of 5.3 newborns per 100 000 live births.

Of the 70 patients, 39 were boys and 31 were girls, with a median age of one day (0–480 days) at presentation (Table 1). In 41 patients (59%) the pulmonary blood flow originated from a single patent ductus arteriosus (PDA) supplying confluent pulmonary arteries (Group 1). In 29 patients (41%) the pulmonary blood flow originated from MAPCAs with or without a contribution from a PDA (Group 2) (Figure 1). No central pulmonary arteries could be found in four patients (5.7%) (Group 2). Various syndromes or chromosomal abnormalities (excluding 22q11 microdeletion syndrome) were found in 10 patients, and 36 patients (51%) had an extracardiac disease (Table 2). The 22q11 microdeletion syndrome was found in five out of the 41 patients without MAPCAs (12%) and in eleven of the 29 patients with MAPCAs.
The various extracardiac diseases are listed in Table 2. Additional cardiac diagnoses are seen in Table 3. The surgical procedures used in all patients are given in Figures 2 and 3. Preceding the surgical corrections in patients without MAPCAs, 31 patients underwent a modified Blalock-Taussig shunt as the first operation, nine patients a central shunt, and one patient underwent a Potts anastomosis (Figure 2). In patients with MAPCAs, there was considerable variation of the type of surgery performed both initially and at subsequent procedures (Figure 3). Unifocalisation of the MAPCAs was performed in 19 of these patients, seven of whom underwent unifocalisation in combination with a Blalock-Taussig shunt and three in combination with a central shunt at first surgery. In four patients, unifocalisation was achieved together with correction at the first surgery. Three patients underwent a Sano shunt and in six patients a transannular patch was inserted. Corrective surgery was accomplished in 58 of the 70 patients (83%) at a median age of 16 months (4 days–19 years). The numbers of patients in Groups 1 and 2 receiving corrective surgery were 35/41 (85%) and 23/29 (79%), respectively (n.s.). Fenestration of the patch to close the VSD was used in two patients in Group 1 and in nine in Group 2 (p < 0.02). Both patients in group 1 who had fenestrated VSD patches were alive at follow-up. In one patient, the homograft was anastomosed only to the left pulmonary artery due to interruption of the right pulmonary artery after a complication following a Blalock-Taussig shunt operation; this patient had acceptable systemic right ventricular pressure. In the other patient, the fenestration had closed spontaneously. Among the nine patients with PA-VSD and MAPCAs who received a fenestrated VSD patch three died: one who had Holt-Oram syndrome and a bronchial stenosis and concomitant pneumonia, one due to an infection, and one after fatal lung bleeding with cardiac arrest at catheterisation. These three patients all had a systemic right ventricular pressure at follow-up before death. Among the remaining six patients with a fenestrated VSD patch, all have a right ventricular pressure lower than the systemic pressure with a left to right shunt across the fenestration not considered to indicate closure of the fenestration. In one of these six patients the fenestration even closed spontaneously. Estimating the residual right ventricular pressure in survivors after corrective surgery by using Doppler measurements at follow-up, we found 21/31 (67%) of the patients without MAPCAs and 9/18 (50%) of patients with MAPCAs (n.s) to have a right ventricular pressure lower than half the systemic pressure.

In Group 1, 18 of the 41 patients had undergone one conduit exchange during the study period, and one of these had undergone two conduit exchanges. In Group 2, five patients out of 29 had undergone a conduit exchange, two of whom patients had undergone

| TABLE 1  | Clinical and demographic data for the PA-VSD patients included in this study |
|-----------|-----------------------------|-----------------------------|-----------------------------|-----------------------------|
| All       | Group 1 (non-MAPCA)         | Group 2 (MAPCA)             | p=value                     |
| N = 70    | n = 41                      | n = 29                      |
| Male      | 39 (66%)                    | 12 (41%)                    | n.s.                        |
| Female    | 31 (34%)                    | 17 (59%)                    | n.s.                        |
| Birthweight, kg, mean (range) | 3.1 (1.4–4.9)               | 3.1 (1.4–4.9)               | n.s.                        |
| Median age at presentation, days (range) | 1 (1–480)                   | 1 (0–480)                   | p < 0.05                    |
| Median age at first operation, days (range) | 12 (1 day–6.9 years)        | 7 (1 day–148)               | 56 (2 days–6.9 years)       | p < 0.001                    |
| Intrauterine diagnosis* | 7                          | 1                           | 6                           | n.s.                        |

Group 1: PA-VSD only, Group 2: PA-VSD+MAPCAs.

*Intrauterine diagnosis only from 2011.

**FIGURE 1** Pulmonary blood flow in 70 patients with PA-VSD

(38%, p < 0.05). The various extracardiac diseases are listed in Table 2. Additional cardiac diagnoses are seen in Table 3.
two exchanges. Catheter interventions, mostly balloon dilatation with or without stent insertion due to pulmonary artery branch steno-
sis, were performed in 37 patients, 19 in Group 1 and in 18 in
Group 2 (Table 4). A Melody® valve was inserted in eight patients:
six in Group 1 and two in Group 2 (Figures 2 and 3). One patient in
Group 1 underwent two Melody® valve insertions.

Death occurred in 18 patients, at a median age of 1.3 years
(9 days–19 years). Three died within 30 days of the last major sur-
gery, and 15 later than this (Table 5). Ten patients, six in Group 1
(14.6%) and four in Group 2 (13.8%), died before corrective surgery
could be undertaken. Eight patients died after corrective surgery,
four in each group. Survival at follow-up was similar in patients with
and without MAPCAs (72.4% vs. 75.6%, n.s); the median follow-up
time was 14.3 years (3.2–41.8) (Figure 4).

All deaths except three were related to cardiac disease: one
death was the result of a car accident, one due to a lethal intracranial
complication after an episode of otitis, and the third due to severe
scoliosis and pronounced respiratory insufficiency (Table 4). We
found no statistically significant difference in mortality when com-
paring patients with (10 patients, 29%) and without (8 patients, 22%)
any syndrome or extracardiac disease (Table 4).

4 | DISCUSSION
The present study describes the outcome of 70 patients treated
for PA-VSD at one hospital in Sweden from a very uniform and
constant referral area, with no private alternative for treatment,
thereby comprising an unselected cohort of patients. There were no drop-outs, thus ensuring complete and reliable data on follow-up. The incidence of 5.3 patients with PA-VSD per 100 000 live births in the present study is well in line with previous reports between 4.2 and 10 per 100 000 live births.\textsuperscript{1–3,17,18} When grouping the patients into those with PA-VSD only and those with MAPCAs we found a similar distribution, that is 59% and 41%, respectively. This is in agreement with a study by Kaskinen et al\textsuperscript{2} who reported the long-term outcome in patients with PA-VSD born in Finland 1970–2007. In patients with both PA-VSD and MAPCAs (Group 2), four (13.8%) lacked central pulmonary arteries (Figure 1), well in line with previous reports.\textsuperscript{1,19}
The significantly earlier clinical presentation in our patients with PA-VSD where the pulmonary blood flow was solely dependent on ductal flow, compared to those with MAPCA connections, was clear in our study. This has been described in some other reports, while others, surprisingly, have not found such a difference. In spite of studying a rather large and population-based cohort, there were no differences found, in contrast to other studies. This discrepancy is likely due to the uneven introduction of screening programmes in different regions. In a comprehensive register study on live-borns with major congenital heart disease in Denmark during the period 1996–2013, the number of newborns with PA-VSD was found to decline significantly as a consequence of the introduction of generalised prenatal screening starting in 2004. Early information on the long-term outcome in patients with this complex lesion is of the utmost importance to the foetal cardiologist to be able to give relevant advice.

A main finding of the present study was a relatively high survival rate (75%) at long-term follow-up, with no statistically significant difference between patients with or without MAPCAs. This finding is in line with the results reported in other studies. However, any study on the outcome of patients with PA-VSD should clearly separate the results into the groups according to the type of pulmonary blood supply, as the surgical strategy and surgical options vary greatly between those with and without MAPCAs.

The surgical strategy at our hospital for patients in whom the pulmonary blood flow supplied by a PDA with no MAPCAs is to perform corrective surgery after an initial shunt operation, either using a modified Blalock–Taussig shunt or a central shunt (75% vs. 22%, Figure 2). Death after these procedures occurred in six patients (15%): five after having received a Blalock–Taussig shunt and one after surgery to insert a central shunt. This confirms the findings of other studies of relatively high mortality and morbidity rates after shunt operations. This has promoted the development of either right ventricle to pulmonary artery shunt operations, or other types of palliative right ventricle to pulmonary artery connection. The cause of death after palliative shunt surgery in this study varied, including difficulties in creating an increased pulmonary blood flow due to diminutive pulmonary arteries, shunt dysfunction combined with infections, while in one case pulmonary oedema contributed to the fatal outcome. Corrective surgery was accomplished in 35/41 (85%) of the patients without MAPCAs and 23/29 (72%) with MAPCAs (Table 4). These are considerably higher than the values presented by Kaskinen et al, who studied all patients in Finland with PA-VSD from 1970 to 2007, where corrective surgery was performed in 42/66 (64%) of patients without MAPCAs and in 12/43 (28%) of those with MAPCAs. In a comprehensive study carried out in Toronto, Canada, also covering a long study period, the proportions of patients receiving corrective surgery were closer to the findings of the present study. Four patients (8.8%) with PA-VSD without MAPCAs in the present study died after corrective surgery, two due to complications after the second corrective surgery (conduit exchanges), while the other two deaths were sudden and unexpected; one of these patients also had severe scoliosis and pronounced respiratory insufficiency.

Most recent reports concerning patients with PA-VSD have, however, dealt with the patient group in which MAPCAs contribute to the pulmonary blood flow, and various surgical strategies have...
been developed based on extensive clinical research and experience. The initial surgical strategy for these patients at our hospital has varied considerably, including: a modified Blalock–Taussig shunt with or without concomitant unifocalisation of the MAPCAs, inserting a central aorto-pulmonary shunt in combination with unifocalisation, a right ventricle to pulmonary artery connection using either a Sano shunt or a transannular patch procedure, or by primary correction in combination with unifocalisation (Figure 3). Such diversity in the initial surgical procedures used is hardly surprising considering the relative rarity of PA-VSD and MAPCAs per se, the variability of the MAPCAs configuration in patients, and also the long period covered by the present study. Not only is the care provided customised to the individual patient, but also depends on the surgeon’s preferences. The survival of patients with PA-VSD with MAPCAs in the present study (69%) was in line that reported previously, but did not reach the survival rates reported by others. In spite of the greater difficulty in achieving complete repair in the MAPCAs group in the present study, long-term survival was similar in the two groups, although a significantly higher number of patients with MAPCAs had undergone fenestration of the VSD patch at correction.

As reported in previous studies, associated comorbidity and syndromes were common in patients with PA-VSD, reaching an incidence of almost 50%. However, a statistically significant difference was only found in the incidence of the 22q11 microdeletion syndrome between patients with and without MAPCAs (38% vs. 15%, p < 0.05, Table 2). We could not confirm the finding reported in several previous studies that 22q11 deletion syndrome was associated with increased mortality at surgery for PA-VSD patients. Also, somewhat surprisingly, we found no increased mortality in patients with other extracardiac disease (Table 4).

### 4.1 Limitations

The study is retrospective with medical charts being scrutinised to retrieve relevant clinical data, some of which, for instance data on right ventricular pressure after correction, exercise testing and

| TABLE 4 Corrective surgery and mortality. | All N = 70 | Group 1 (non-MAPCA) n = 41 | Group 2 (MAPCA) n = 29 | p-value |
|------------------------------------------|-----------|--------------------------|------------------------|--------|
| Patients with complete correction (%)   | 58        | 35 (85%)                 | 23 (79%)               | n.s.   |
| Median age at correction, years (range)  | 1.3 (4 days–19 years) | 1.2 (4 days–6.5 years) | 1.8 (10 days–19 years) | n.s.   |
| Number of patients with one conduit exchange | 23        | 18 (44%)                 | 5 (17%)                | p < 0.05 |
| Number of patients with two conduit exchanges | 3         | 1 (2%)                   | 2 (7%)                 | -      |
| Number of patients with Melody insertion | 8         | 6 (15%)                  | 2 (7%)                 | -      |
| Number of patients with two Melody insertions | 1         | 1 (2%)                   | 0                      | -      |
| Total mortality (%)                     | 18 (26%)  | 10 (24%)                 | 8 (27%)                | n.s.   |
| Median age at death, years (range)      | 1.6 (9 days–27.3 years) | 0.8 (9 days–27.3 years) | 3.8 (1.3–19.1 years) | p < 0.01 |
| Mortality: No extracardiac associations | 8 (22%)   | 6 (25%)                  | 2 (16%)                | -      |
| Mortality: Extracardiac associations including 22q11 and other syndromes | 10 (29%) | 4 (24%)                  | 6 (35%)                | n.s.   |
| Mortality: 22q11 microdeletion syndrome only | 5 (31%) | 2 (40%)                  | 3 (27%)                | n.s.   |
| Mortality: Syndromes other than 22q11 microdeletion syndrome | 4 (36%) | 2 (33%)                  | 2 (40%)                | -      |
| Mortality: Extracardiac associations, no verified syndrome | 1 (10%) | 0                        | 1 (100%)               | -      |
| Mortality: Cardiac causes | 15 (83%) | 9 (90%)                  | 6 (75%)                | n.s.   |
| Mortality: Other causes | 3 (17%) | 1 (10%)                  | 2 (25%)                | -      |

Group 1: PA-VSD only, Group 2: PA-VSD+MAPCAs.
| Group and gender (M/F) | Extracardiac associations                                                                 | Survival     | Last type of surgery (number) | Corrected Yes/No (Fenestrated patch Y/N) | Time after last surgery | Cause of death                                                                 |
|------------------------|-------------------------------------------------------------------------------------------|--------------|-------------------------------|------------------------------------------|-------------------------|--------------------------------------------------------------------------------|
| 1 (F)                  | 22q11 microdeletion syndrome, scoliosis, respiratory insufficiency                        | 27.3 years   | Correction (3)                | Y (N)                                    | 22.4 years              | Respiratory insufficiency                                                     |
| 1 (M)                  | Charge syndrome                                                                           | 89 days      | Blalock-Taussig shunt (2)     | N                                        | 25 days                 | Desaturation                                                                  |
| 1 (M)                  | None                                                                                      | 9 days       | Shunt exchange and right    | N                                        | 0                       | Desaturation                                                                  |
| 1 (M)                  | Trisomy 21                                                                                | 1.1 year     | Correction (2)                | Y (N)                                    | 38 days                 | VSD patch dehiscence                                                          |
| 1 (F)                  | 22q11 microdeletion syndrome                                                              | 1.0 year     | Correction (2)                | Y (N)                                    | 11 days                 | Multi-organ failure and myocardial ischaemia after surgery                    |
| 1 (M)                  | None                                                                                      | 198 days     | Blalock-Taussig shunt (1)     | N                                        | 186                     | Pulmonary oedema                                                              |
| 1 (M)                  | None                                                                                      | 12 days      | Blalock-Taussig shunt (1)     | N                                        | 0                       | Arrhythmia after surgery                                                      |
| 1 (F)                  | None                                                                                      | 63 days      | Blalock-Taussig shunt (1)     | N                                        | 22 days                 | Intra cerebral haemorrhage during ECMO                                        |
| 1 (M)                  | None                                                                                      | 14.8 years   | Conduit exchange (4)          | Y (N)                                    | 4.3 years               | Sudden cardiac death (abroad)                                                 |
| 1 (M)                  | None                                                                                      | 1.0 year     | Blalock-Taussig shunt (1)     | N                                        | 1.0 years               | Gastroenteritis with arrhythmia as a complication                             |
| 2 (F)                  | Congenital kyphosis                                                                        | 19.1 years   | Correction (5)                | Y (N)                                    | 2.3 years               | VSD patch dehiscence                                                          |
| 2 (F)                  | 22q11 microdeletion syndrome                                                              | 7 years      | Correction (3)                | Y (N)                                    | 4.2 years               | Car accident                                                                  |
| 2 (F)                  | Holt-Oram syndrome                                                                        | 9.6 years    | Correction (5)                | Y (Y)                                    | 4.9 years               | Heart failure                                                                 |
| 2 (M)                  | 22q11 microdeletion syndrome                                                              | 3.7 years    | Correction (2)                | Y (Y)                                    | 2.2 years               | Pulmonary bleeding due to catheter complication                               |
| 2 (F)                  | Chromosome 18P deletion                                                                   | 1.8 years    | Blalock-Taussig shunt exchange (2) | N                                        | 45 days                 | Shunt thrombus and concomitant viral infection                                |
| 2 (F)                  | None                                                                                      | 7.5 years    | Blalock-Taussig shunt (1)     | N                                        | 7.4 years               | Otitis with bacterial emboli to the brain                                    |
| 2 (M)                  | 22q11 microdeletion syndrome                                                              | 2.8 years    | Correction (2)                | Y (Y)                                    | 1.0 years               | Shunt perforation during catheterisation                                     |
| 2 (M)                  | None                                                                                      | 1.3 years    | Central shunt (3)             | N                                        | 124 days                | Shunt thrombus after catheterisation                                         |

* Group 1: PA-VSD only, Group 2: PA-VSD+MAPCAs.
quality of life, are incomplete rendering the evaluation of the long-term outcome apart from mortality, type of interventions and comorbidities difficult. The inclusion of a smaller cohort with their first surgery even before 1994 may to some extent skew some of the grouped data.

5 | CONCLUSIONS

Pulmonary atresia and ventricular septal defect with or without MAPCAs is a rare malformation; the incidence of patients receiving surgery in this complete cohort from a very stable referral area being 5.3% per 100 000 newborns. No difference was seen in survival between the groups according to Kaplan–Meier analysis (Figure 4). Although patients with MAPCAs required more surgical interventions, on average, than the group without MAPCAs, long-term survival was similar in the two groups, (72.4% vs 75.6%, n.s.) after a median follow-up time of 14.3 years (3.2–41.8 years). Associated comorbidities and syndromes were common in patients with PA-VSD both with and without MAPCAs in total, almost 50%. No significant difference was found in survival between patients with and without 22q11 microdeletion syndrome, or with and without comorbidities and syndromes in general.

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CONFLICT OF INTERESTS

The authors declare no conflicts of interests.

ORCID

Erik Wiezell  https://orcid.org/0000-0003-4394-8427
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