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Current outcomes of live-born children with double outlet right ventricle in Norway

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Key question

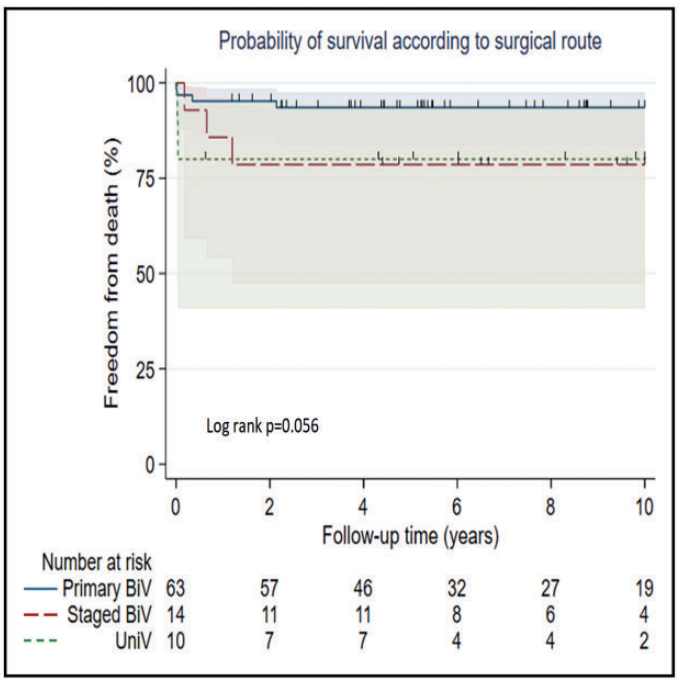
What is the outcome including complications, re-interventions and mortality for live-born children with DORV in Norway?

Key finding(s)

Children with DORV receive tailored treatment comprising primary or staged BiV or UniV repair with favorable outcomes.

Take-home message

Despite low rates of mortality and severe complications, AVSD is a risk factor for death to be accounted for in tailored treatment of DORV.



Abstract

OBJECTIVES: This population-based, comprehensive, retrospective study presented the clinical outcomes of all children born in Norway between 2003 and 2017 with double outlet right ventricle (DORV).

METHODS: All children born with DORV between 2003 and 2017 were identified in the Oslo University Hospital registry. Patients' characteristics, interventions, complications and deaths were recorded. Echocardiographic data were reviewed for classification according to current standards. We investigated time-dependent surgical reintervention and mortality using Kaplan–Meier analyses and determinants of treatment complications, reintervention and death using regression analyses.

RESULTS: Ninety-three children with DORV represented an annual median prevalence of 1.18 per 10 000 births in Norway. Six children received palliative care. With an intention to treat, a surgical route with the primary biventricular repair was followed for 62 children, staged biventricular repair for 15 and univentricular repair for 10 children. Major complications occurred in 1.0% and 6.2% of children

following catheter or surgical intervention, respectively. No significant determinants of the complications were identified. Overall survival following treatment was 91.9%, 90.8%, 89.5% and 89.5% and corresponding freedom from surgical reintervention was 88.0%, 79.0%, 74.9% and 69.4% at 1, 2, 5 and 10 years, respectively. The presence of atrioventricular septal defect predicted an increased risk of mortality (hazard ratio: 7.16) but did not increase the risk of surgical reintervention.

CONCLUSIONS: In Norway, most children receive tailored treatment for DORV with low rates of complications, surgical reinterventions and mortality. However, atrioventricular septal defect remains a potential determinant of postoperative death.

Keywords: Double outlet right ventricle • DORV • Biventricular • Univentricular • Complication • Reintervention

ABBREVIATIONS

AVSD	Atrioventricular septal defect
BiV	Biventricular
CI	Confidence interval
DORV	Double outlet right ventricle
ICU	Intensive care unit
IQRs	Interquartile ranges
nc-VSD	Non-committed ventricular septal defect
RV	Right ventricle
STS	Society of Thoracic Surgeons
TGAs	Transposition of the great arteries
TOF	Tetralogy of Fallot
UniV	Univentricular
VSD	Ventricular septal defect

INTRODUCTION

Double outlet right ventricle (DORV) is a complex congenital heart disease that represents a multitude of morphological variants of the basic concept of both great arteries arising primarily from the right ventricle (RV) [1, 2]. The definition of DORV and the distinction between DORV and morphologically bordering cardiac malformations such as transposition of the great arteries (TGAs), tetralogy of Fallot (TOF) and simple ventricular septal defect (VSD) have been debated challenging the comparability of individual studies of this malformation [1–3]. Recently, the classification of the Society of Thoracic Surgeons (STS)-European Association for Cardiothoracic Surgery International Nomenclature and the Association for European Paediatric Cardiology, which defines 4 distinct types of DORV, has been applied in studies of DORV and has thus provided a basis for the comparison of results [4–6]. However, it has been argued that this classification system may be insufficient for planning surgical strategies [7]. An alternative method for describing the morphology in detail has recently been suggested, based on a series of 'essential modifiers' [8].

Owing to the highly variable morphology of DORV, the options for surgical treatment are diverse and encompass both biventricular (BiV) and univentricular (UniV) solutions. The choice of surgical route differs greatly in reported series, with BiV repair ranging from 37% to 72% and UniV repair from 23% to 58% [4, 7, 9, 10]. It has been argued that the distinction between BiV and UniV solutions is particularly important in children with inadequate left-sided structures and non-subaortic VSDs. This applies to Rastelli-type repair with an RV-to-pulmonary artery conduit, which has been shown to be associated with higher late mortality and reintervention rates compared to UniV repair [9]. For BiV repair of DORV with complex anatomy, acceptable outcomes can be obtained with both primary and staged approaches [11]. Owing to the divergence in surgical solutions, there are large variations in reported rates of mortality ranging from 4.5% to 14% [4, 5, 10–12].

Few studies have reported on complications following DORV treatment. Two studies have reported rates of 1–24% for various postoperative complications [13, 14], and another study reported a high cumulative incidence (42%) of postoperative complications [15]. We are not aware of reports on complications following catheter interventions or complications registered according to the International Paediatric and Congenital Cardiac Code nomenclature. Furthermore, European studies on DORV outcomes are scarce and mostly represent selected patient series [16–18].

In the present study, we estimated the annual incidence of DORV in Norway over a recent 15-year period and reviewed the outcomes from the time of birth including detailed patients' characteristics, surgical and catheter-based interventions and outcomes encompassing complications, surgical reinterventions and mortality.

MATERIALS AND METHODS

Ethical statement

This study was categorized as internal quality assurance and approved by the local data protection officer, Oslo University Hospital (2 January 2013; number: 2012/18706).

The study was performed at the Department of Paediatric Cardiology, Rikshospitalet, Oslo University Hospital (National Center for Congenital Heart Disease, Norway). Using the database for congenital heart disease (BERTE [19]), all children born between 1 January 2003 and 31 December 2017 diagnosed with DORV, TOF or TGA regardless or additional defects and unbalanced ventricles were identified. Neonatal preoperative echocardiograms were reviewed to confirm the diagnosis of DORV according to the STS criteria defining 4 DORV groups: VSD-type, Fallot-type, TGA-type and non-committed VSD (nc-VSD)-type DORV [20, 21]. Regarding the position of the great arteries, we used the 90% rule [22] applied by Pang *et al.* [7], where 100% of 1 great artery and 90% of the other arose from the RV. TGA-type DORV was defined as cases in which the pulmonary artery overrode the VSD by 50% or more [7]. The distinction between committed and nc-VSD was based on a comparison of the distance between the superior margin of the VSD and the nearest edge of the arterial valves. Cases where the distance was greater than the aortic diameter were classified as non-committed and cases where the distance was equal to or less than the aortic diameter were classified as committed [7].

Baseline characteristics (date of birth and death, sex, gestational age, weight and length at birth and at interventions, syndromes and

non-cardiac malformations) were recorded. Early death was defined as death within 30 days of surgery or death before discharge following surgery, and late and very late death as death between 30–365 and 365 days after surgery for discharged patients, respectively. Furthermore, cross-clamp duration, postoperative days on ventilator, postoperative days in the intensive care unit and postoperative length of stay were recorded. Data on catheter-based interventions and complications were extracted from a local catheter intervention database [23]. All surgical interventions and complications were retrospectively registered. All interventions, complications and deaths until 31 June 2019 were included. Interventions and complications were classified according to the International Paediatric and Congenital Cardiac Code classification system [23, 24] using 5 grades ranging from 1 to 5 (1 = none, 2 = minor, 3 = moderate, 4 = major and 5 = catastrophic severity). Based on the course of surgical intervention, all children were categorized as receiving either primary or staged BiV or UniV repair. As reported by Oladunjoye *et al.* [11], primary BiV repair encompasses complete anatomic repair of DORV without prior palliative surgery except pulmonary banding. Staged BiV repair included cases in which a sequence of separate surgical procedures commencing with palliative procedures, except pulmonary banding, was initiated with the aim of complete BiV repair. UniV repair included cases in which circulation with 1 functional ventricle was established. The annual rates of DORV were calculated using the official annual birth statistics in Norway obtained from the National Database of Statistics Norway [25].

Statistical analysis

Variables are presented as numbers, proportions, medians and interquartile ranges (IQRs). Differences in variables between surgical routes and DORV categories were calculated using nonparametric tests including Kruskal–Wallis, Chi-squared and Fisher's exact tests as appropriate. Significance levels were set at 5%. Logistic regression analyses were applied to identify factors associated with surgical complications. Corresponding analyses for catheterization complications were not performed because of low incidence. Time-dependent surgical reintervention and mortality rates were assessed using Kaplan–Meier analyses. Cox proportional hazards regression analyses were applied to identify determinants for death and competing risks regression analyses for surgical reintervention. The following independent variables were *a priori* included in the regression analyses of surgical complications, mortality and reintervention: sex, DORV type, presence of atrioventricular septal defect (AVSD), presence of syndromes or non-cardiac malformations, surgical route and age at intervention. Because of the low sample size and the risk of type II error, only descriptive univariable regression analyses were performed.

RESULTS

We identified 117 children born between 2003 and 2017 with DORV in the database. After reviewing the pre-interventional echocardiograms, 92 children with DORV fulfilled the predefined inclusion criteria. Thus, 24 children were reclassified accordingly: 9 with TOF, 1 with TGA and 14 with complex congenital heart disease other than DORV. We also reviewed all cases registered as TGA and TOF in the same period, but only 1 child was reclassified as having DORV. The final population-based sample encompassed 93 children. Based on national birthrates, the median annual (IQR) incidence (per 10 000 newborns) of DORV was 1.18 (0.78–1.36). Additional non-cardiac malformations or syndromes were present in 25 (27%) of the children. Of 93 included children, 87 (94%) were given treatment courses with an intention to treat, while 6 (6.5%) children (4 with VSD-type DORV, 2 with Fallot-type DORV) received comfort care because of severe comorbidities resulting from pervasive syndromes (Fig. 1). The surgical strategy was primary BiV repair in 63 (72.4%) children, staged BiV repair in 14 (16.1%) and UniV repair in 10 (11.5%) children, and the median (IQR) follow-up time was 6.4 (3.8–10.8) years. No children were lost to follow-up. No differences were found in the occurrence of catheter or surgical complications, surgical reinterventions or death between children who underwent primary BiV, staged BiV or UniV repair.

Characteristics, interventions and outcomes of the 87 treated patients are shown in Table 1. DORV types included 18 (21%) children with VSD type, 24 (28%) with Fallot type, 37 (43%) with TGA type and 8 (9.2%) with nc-VSD-type DORV. Six (6.9%) patients had complete AVSD, all of whom had Fallot-type DORV. Additional non-cardiac malformations or syndromes were present in 19 (22%) of the treated children, with higher prevalence among children with VSD- and Fallot-type DORV. The median (IQR) of the patients' age at first surgery was 1.3 (0.6–16) months with the lowest ages among children with TGA-type and VSD-type DORV. Eight children received pulmonary banding as the first surgical intervention at a median (IQR) age of 0.8 (0.7–1.2) months. The median (IQR) of patients' age at final repair was 4.7 (0.7–35) months with increased differences in ages between DORV types with early repair (TGA type and VSD type) and late repair (Fallot type and nc-VSD type). A total of 101 catheter-based procedures were performed in the 87 treated children. Of the catheter procedures, 54 (53%) were diagnostic in 34 children and 47 (47%) were interventional in 29 children with pulmonary angioplasty constituting most of the interventional procedures. Children with VSD-type DORV had fewer catheter procedures than those in other DORV groups. Of 145 surgical procedures, 92 (63%) were BiV, 24 (13%) UniV and 30 (21%) reinterventions with no differences in distribution between the DORV types.

A total of 5 (5.0%) complications occurred following catheter-based procedures (Supplementary Material, Table S1). There were no deaths, but 2 cases were categorized as grade 3 (moderate) and 1 as grade 4 (major) complications. Overall, 32% of the surgical interventions (47 of 145) were followed by complications in 38 children. Four of these children died, 5 had major complications and 37 were classified as moderate. Complications following catheterization were attributable to device (40%) or intervention (20%), haemodynamics (20%), arrhythmia (20%) and respiratory factors (20%) (Supplementary Material, Table S2). Surgical complications were attributable to respiratory (45%), haemodynamic (28%), arrhythmia (11%), infection (6.4%), surgical (6.4%) and neurological (4.3%) factors (Supplementary Material, Table S2). Twenty-four surgical complications occurred among children who underwent primary BiV repair, 12 among children who underwent staged BiV repair and 11 among children who underwent UniV repair ($P = 0.3$). Logistic regression analyses did not reveal any significant determinants of surgical complications (data not shown).

The median (IQR) cross-clamp time was 83 (51–120) min with VSD- and Fallot-type DORV having lower median cross-clamp times (60 and 59 min, respectively) than those found in TGA- and

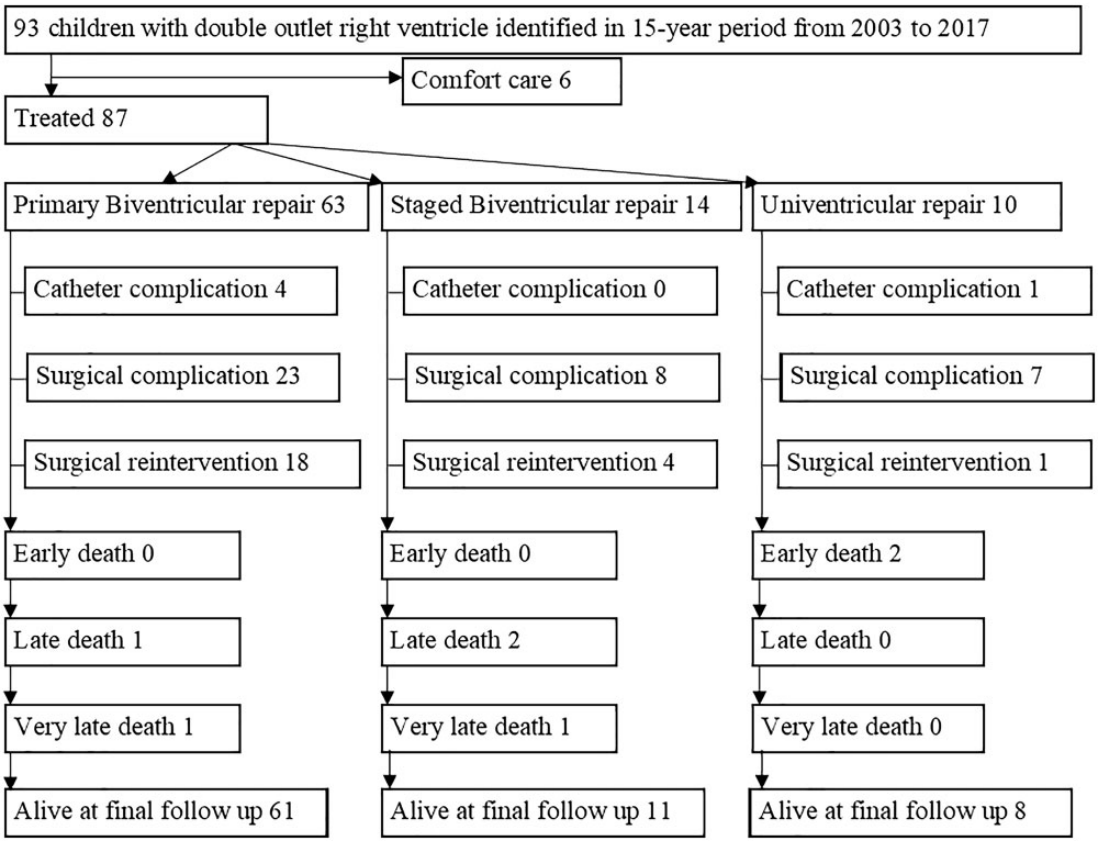


Figure 1: Flow chart: identification of children born with double outlet right ventricle in the BERTE register at the National Center for Congenital Heart Disease in Norway.

nc-VSD-type DORV (107 and 110 min, respectively). The median (IQR) number of postoperative days on ventilator, in ICU and for hospital stay were 2 (1-4), 7 (4–10) and 15 (9–24) days, respectively. No significant differences were found between DORV-type groups for these variables. The number of deaths among all treated children was 9 (10%) with no significant differences between DORV groups. Four (4.6%) deaths occurred within 30 days of surgery and were classified as grade 5 complications. Three (3.4%) deaths occurred between 30 days and 1 year after surgery and were classified as late. Very late deaths (>1 year postoperatively) occurred in 2 (2.3%) children. The estimated overall survival was 91.9% at 1 year, 90.8% at 2 years and 89.5% at 5 and 10 years for all children treated for DORV (Table 2).

No significant differences in survival were found between children undergoing primary BiV, staged BiV or UniV repair, although a trend towards a higher probability of survival following primary BiV repair was seen (Fig. 2). Children with AVSD had a significantly higher risk of mortality (Fig. 3). Estimated overall freedom from surgical reintervention was 88.0%, 79.0%, 74.9% and 69.4% at 1, 2, 5 and 10 years for the treated children (Table 2). No differences in freedom from surgical reintervention were found between the routes of surgical repair and the presence of AVSD. In univariable Cox proportional hazards analyses (Supplementary Material, Table S3), the presence of AVSD was the only determinant of increased risk of death (hazard ratio: 7.16, confidence interval: 1.78–28.8). No determinants were identified for the risk of surgical reintervention in competing risks analyses (Supplementary Material, Table S3).

DISCUSSION

In this retrospective, population-based study of DORV in Norway during a recent 15-year period, we found a median annual cumulative incidence of 1.18 per 10 000 live births and a total mortality of 16% including death following palliative care. TGA-type DORV was the most common variant. The far majority of the children were offered tailored treatment with either primary BiV, staged BiV or UniV repair with favourable outcomes. Most complications following catheter-based or surgical interventions were of low severity and without permanent adverse consequences. The presence of AVSD was the only potential determinant of increased risk of death.

Our finding of a median annual cumulative incidence of 1.18 per 10 000 live births is in line with previous reports of DORV occurrence. Summarizing data from 16 studies before year 2000, Hoffman and Kaplan [26] reported an average incidence of 1.05 of DORV. More recent nationwide studies of congenital heart defects in the USA and in Taiwan reported slightly higher incidences of DORV (1.7 per 10 000 in the USA, 1.5 per 10 000 in Taiwan) [27, 28]. In a large registry-based nationwide study of congenital heart defects in Norway, Leirgul *et al.* [29] found a lower incidence of DORV among live-born children (0.7 per 10 000). Discrepancies in reported incidences may be explained by differences in diagnostic definitions of DORV and thoroughness of the case review. In our study, we reclassified 25 children in the BERTE database after reviewing preoperative neonatal echocardiograms and applying the described diagnostic criteria.

Table 1: Characteristics for 87 treated children with double outlet right ventricle

DORV type	DORV all	VSD type	Fallot type	TGA type	nc-VSD type	*P*-Value*
Total, *n* (%)	87 (100)	18 (21)	24 (28)	37 (43)	8 (9.2)	
Sex						0.3
Females	25 (29)	4 (4.6)	10 (11)	8 (9.2)	3 (3.5)	
Males	62 (71)	14 (16)	14 (16)	29 (33)	5 (5.8)	
Birth weight (kg)[a]	3.4 (2.8–4.0)	3.0 (2.8–3.7)	2.9 (2.3–3.2)	3.7 (3.4–4.2)	3.7 (2.8–4.1)	0.002*
Syndrome/malformation	19 (22)	7 (8.1)	8 (9.2)	3 (3.5)	1 (1.2)	0.02
AVSD	6 (6.9)	0 (0)	6 (6.9)	0 (0)	0 (0)	0.001
Doubly committed VSD	1 (1.2)	0 (0)	1 (1.2)	0 (0)	0 (0)	0.6
Age at 1st surgery (months)	1.3 (0.6–16)	2.7 (1–5.4)	17 (1.8–33)	0.6 (0.4–0.8)	21 (0.8–63)	0.0001*
Age at repair (months)	4.7 (0.7–35)	2.7 (0.9–6.3)	28 (13–46)	0.7 (0.4–28)	59 (24–156)	0.0001*
Patients surgical route						
Primary BiV repair	63 (72)	17 (20)	13 (15)	30 (34)	3 (3.4)	0.007
Staged BiV repair	14 (16)	0 (0)	8 (9.2)	3 (3.4)	3 (3.4)	0.01
UniV repair	10 (11)	1 (1.2)	3 (3.4)	4 (4.6)	2 (2.3)	0.5
Pulmonary banding	8 (9.2)	2 (2.3)	2 (2.3)	2 (2.3)	2 (2.3)	0.3
Cross-clamp time (min)	83 (51–120)	60 (36–115)	59 (44–80)	107 (74–134)	110 (54–124)	0.007*
Ventilation time (days)	2 (1–4)	1.5 (1–5)	1.5 (1–4)	2 (1.5–4)	1.5 (1–6)	0.7*
ICU stay (days)	7 (4–10)	5.5 (2–13)	7 (4–9)	7 (4–9.5)	6 (3–12)	1.0*
Hospital stay (days)	15 (9–24)	18 (7–26)	20 (11–26)	14 (10–24)	15 (8.5–18)	0.7*
Deaths, *n* (%)	9 (10)	3 (3.5)	2 (2.3)	2 (2.3)	2 (2.3)	0.1
Early (<30 days)	4 (4.6)	2 (2.3)	0 (0)	1 (1.2)	1 (1.2)	0.2
Late (≥30 days)	3 (3.4)	1 (1.2)	0 (0)	1 (1.2)	1 (1.2)	0.1
Very late (>1 year)	2 (2.3)	0 (0)	2 (2.3)	0 (0)	0 (0)	0.2
Catheter procedures	101 (100)	8 (7.9)	32 (32)	52 (51)	9 (8.9)	0.02
Diagnostic	54 (53)	7 (6.9)	15 (15)	24 (24)	8 (7.9)	0.1
Interventional	47 (47)	1 (1.0)	17 (17)	28 (28)	1 (1.0)	0.2
Catheter complications	5 (5.0)	0 (0)	1 (1.0)	4 (4.0)	0 (0)	0.2
Surgical procedures	145 (100)	26 (18)	40 (28)	64 (44)	15 (10)	0.4
Biventricular	92 (63)	18 (12)	28 (19)	37 (26)	9 (6.2)	0.6
Univentricular	23 (16)	2 (1.4)	7 (4.8)	10 (6.9)	4 (2.8)	0.5
Reintervention	30 (21)	6 (4.1)	5 (3.5)	17 (12)	2 (1.4)	0.3
Surgical complications	47 (32)	9 (6.2)	13 (9.0)	21 (14)	4 (2.8)	1.0

Variables presented as *n* (%) or medians (interquartile range). *P*-values refer to tests for differences between DORV groups.

[a]Birth weights missing for 5 children.

*Kruskal–Wallis, else Fisher's exact test.

AVSD: atrioventricular septal defect; BiV: biventricular; DORV: double outlet right ventricle; ICU: intensive care unit; nc-VSD: non-committed ventricular septal defect; TGAs: transposition of the great arteries; UniV: univentricular; VSD: ventricular septal defect.

Table 2: Kaplan–Meier models for freedom from death or surgical reintervention in 87 treated children with double outlet right ventricle

	% at 1 year (CI)	% at 2 years (CI)	% at 5 years (CI)	% at 10 years (CI)
Freedom from death				
All DORV	91.9 (83.8–96.1)	90.8 (82.4–95.3)	89.5 (80.8–94.4)	89.5 (80.8–94.4)
Primary BiV repair	95.2 (86.0–98.4)	95.2 (86.0–98.4)	93.5 (83.7–97.5)	93.5 (83.7–97.5)
Staged BiV repair	85.7 (53.9–96.2)	78.6 (47.3–92.5)	78.6 (47.3–92.5)	78.6 (47.3–92.5)
UniV repair	80.0 (40.9–94.6)	80.0 (40.9–94.6)	80.0 (40.9–94.6)	80.0 (40.9–94.6)
No AVSD	92.6 (84.3–96.6)	92.6 (84.3–96.6)	92.6 (84.3–96.6)	92.6 (84.3–96.6)
Presence of AVSD	83.3 (27.3–97.5)	62.5 (14.2–89.3)	41.7 (5.60–76.7)	41.7 (5.60–76.7)
Freedom from surgical reintervention				
All DORV	88.0 (78.8–93.4)	79.0 (68.4–86.4)	74.9 (63.8–83.1)	69.4 (56.1–79.4)
Primary BiV repair	88.4 (77.1–94.3)	76.4 (63.4–85.3)	74.7 (61.5–83.9)	67.0 (50.5–79.0)
Staged BiV repair	85.7 (53.9–96.2)	85.7 (53.9–96.2)	68.6 (35.9–87.0)	68.6 (35.9–87.0)
UniV repair	90.0 (47.3–98.5)	90.0 (47.3–98.5)	90.0 (47.3–98.5)	90.0 (47.3–98.5)
No AVSD	88.4 (78.8–93.8)	78.9 (67.9–86.5)	74.6 (63.1–83.0)	68.8 (55.1–79.2)
Presence of AVSD	83.3 (27.3–97.5)	83.3 (27.3–97.5)	83.3 (27.3–97.5)	83.3 (27.3–97.5)

AVSD: atrioventricular septal defect; BiV: biventricular; CI: confidence interval; DORV: double outlet right ventricle; UniV: univentricular.

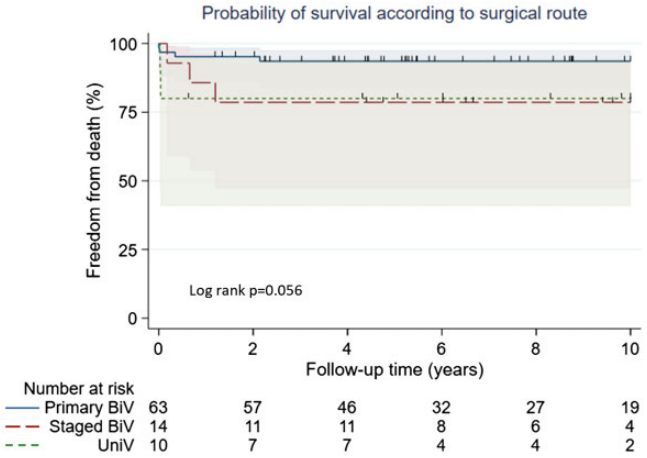


Figure 2: Probability of survival for children with double outlet right ventricle according to surgical route with 95% confidence intervals. BiV: biventricular repair; UniV: univentricular repair.

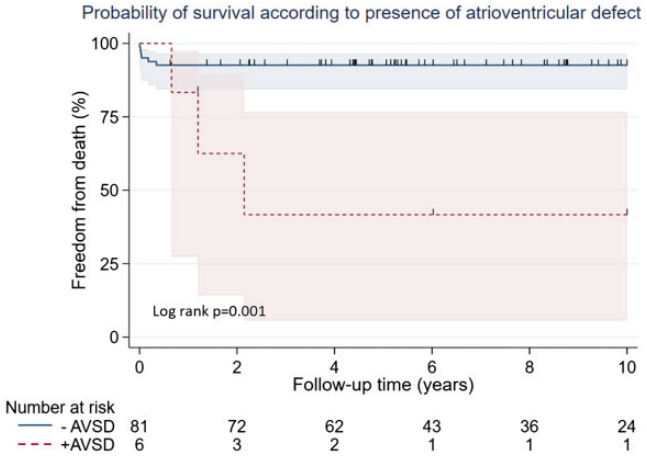


Figure 3: Probability of survival for children with double outlet right ventricle according to the presence of atrioventricular septal defect with 95% confidence intervals. AVSD: atrioventricular septal defect.

The results of the distribution of DORV types in our study with TGA-type DORV being most frequent and nc-VSD type being present in <10% differ from the results of similar studies using STS criteria. In a study of 50 children who underwent surgical repair for DORV, Artrip *et al.* [30] reported Fallot-type DORV to be the most frequent and TGA type as the least frequent. In a larger study of children undergoing BiV repair for DORV, Meng *et al.* [6] reported an almost even distribution between the 4 DORV types with a slightly higher proportion of TGA-type DORV at 27%. However, both studies included selected cohorts that underwent surgical intervention for DORV and may not be readily comparable to our unselected population.

Owing to the highly variable morphology of DORV, the surgical treatment approach is broad and encompasses both BiV and UniV solutions. The choice of surgical route differs greatly in the reported study series with BiV repair ranging from 37% to 72% and UniV repair from 23% to 58% [4, 7, 9, 10]. Most children in our study underwent primary BiV repair, while staged BiV repair (17%) and UniV repair (11%) were less commonly used. It has been argued that the distinction between candidates for BiV and UniV repair should be made in DORV cases with inadequate left-sided structures and non-subaortic VSDs. This applies particularly to Rastelli-type repair with an RV-to-pulmonary artery conduit, which has been shown to be associated with higher rates of late mortality and reintervention than those in UniV repair [9].

In accordance with the heterogeneity of reported DORV cohorts, outcomes vary between studies and rates of mortality ranging from 6% to 14% have been published [4, 5, 10–12]. Recently, acceptable outcomes were reported for both primary and staged approaches for BiV repair of DORV with complex anatomy, with overall survival of 89% at 5 years after surgery [11]. The children selected for primary repair probably represent a less complicated group, and the lack of a statistical difference in mortality between surgical routes in our study may be explained by insufficient statistical power. This may also apply to the group of children with DORV without syndromes and non-cardiac malformations. Only AVSD was identified as an independent determinant of postoperative mortality. Three of 6 children with AVSD died during follow-up; however, 2 of these had pervasive syndromes (CHARGE and DiGeorge, respectively). Although non-cardiac malformations or syndromes were not found to be associated with mortality in our study, the size of our cohort could be speculated to result in type II error. Our finding of a cumulative mortality of 10% is mid-range, underscoring the lack of improvement in published results over the last 2 decades. In addition, 6.5% of the children died in palliative care because of severe comorbidity, and the total mortality was significant. The lack of larger population-based studies with a consistent classification system suitable for the classification of diagnosis, treatment and follow-up of children with DORV, including the most complex variants, has been emphasized [8]. The STS classification may be insufficient to achieve a common understanding and improve the treatment of this complex cardiac malformation.

Our study is the first to report complications following catheter-based and surgical interventions for the treatment of DORV. Complications followed 32% of the surgical procedures and 5% of the catheter-based procedures, which was lower than those reported by the only other study on cumulative postoperative complications [15]. The study by Hayes *et al.* [15] included only patients who received BiV repair for Taussig–Bing anomaly (TGA-type DORV), who may be at higher risk. However, there was no difference between the types of DORV in our study. The cumulative incidence of 5% catheter complications corresponds well with the general results of catheter interventions at our institution [23].

We consider population-based nationwide data, as well as the strict classification based on detailed reviews of echocardiographic criteria as the major strengths of this study. However, we cannot completely exclude the possibility that a patient may have died before surgery without being recorded in the registries. The relatively low total number of children limits the statistical power, particularly for subgroup analyses.

CONCLUSION

We found an annual incidence of DORV corresponding to 1.07 per 100.000 live births in Norway and found that the total mortality, including death following palliative care, was significant. However, most children with DORV can be offered tailored treatment encompassing primary BiV, staged BiV or UniV repair with favourable outcomes. The mortality rate among treated children seems to have stabilized at a low level in the last few decades,

and severe complications are rare. The presence of AVSD remains an important determinant of increased risk of postoperative death.

SUPPLEMENTARY MATERIAL

Supplementary material is available at *EJCTS* online.

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Data Availability

The data underlying this article cannot be shared publicly due to policy for internal quality assurance databases at Oslo University Hospital. The data will be shared on reasonable request to the corresponding author.

Author contributions

Mads Holten-Andersen: Conceptualization; Data curation; Formal analysis; Funding acquisition; Investigation; Methodology; Project administration; Resources; Software; Visualization; Writing—original draft; Writing—review & editing. **Matthias Lippert:** Methodology; Writing—original draft; Writing—review & editing. **Henrik Holmstrøm:** Conceptualization; Data curation; Methodology; Writing—original draft; Writing—review & editing. **Henrik Brun:** Methodology; Writing—original draft; Writing—review & editing. **Gaute Døhlen:** Conceptualization; Data curation; Methodology; Project administration; Supervision; Writing—original draft; Writing—review & editing.

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