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Preoperative versus postoperative survival in patients with univentricular heart: a nationwide, retrospective study of patients born in 1990–2015

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ABSTRACT

Objectives Few data exist on mortality among patients with univentricular heart (UVH) before surgery. Our aim was to explore the results of intention to perform surgery by estimating preoperative vs postoperative survival in different UVH subgroups.

Design Retrospective.

Setting Tertiary centre for congenital cardiology and congenital heart surgery.

Participants All 595 Norwegian children with UVH born alive from 1990 to 2015, followed until 31 December 2020.

Results One quarter (151/595; 25.4%) were not operated. Among these, only two survived, and 125/149 (83.9%) died within 1 month. Reasons for not operating were that surgery was not feasible in 31.1%, preoperative complications in 25.2%, general health issues in 23.2% and parental decision in 20.5%. In total, 327/595 (55.0%) died; 283/327 (86.5%) already died during the first 2 years of life. Preoperative survival varied widely among the UVH subgroups, ranging from 40/65 (61.5%) among patients with unbalanced atrioventricular septal defect to 39/42 (92.9%) among patients with double inlet left ventricle. Postoperative survival followed a similar pattern. Postoperative survival among patients with hypoplastic left heart syndrome (HLHS) improved significantly (5-year survival, 42.5% vs 75.3% among patients born in 1990–2002 vs 2003–2015; p<0.0001), but not among non-HLHS patients (65.7% vs 72.6%; p=0.22)—among whom several subgroups had a poor prognosis similar to HLHS. A total of 291/595 patients (48.9%) had Fontan surgery

Conclusions Surgery was refrained in one quarter of the patients, among whom almost all died shortly after birth. Long-term prognosis was largely determined during the first 2 years. There was a strong concordance between preoperative and postoperative survival. HLHS survival was improved, but non-HLHS survival did not change significantly. This study demonstrates the complications and outcomes encountering newborns with UVH at all major stages of preoperative and operative treatment.

STRENGTHS AND LIMITATIONS OF THIS STUDY

⇒ The study is based on an essentially unselected national cohort.
⇒ The patients are followed all the way from birth, and through all major stages of treatment.
⇒ As in all observational studies demonstrating associations, causations can not be implied.
⇒ The univentricular heart diagnostic subgroups are heterogeneous, and standardisation is difficult.

INTRODUCTION

Children born with functionally univentricular heart (UVH) are typically managed by a staged surgical approach aimed at culminating in the establishment of Fontan circulation. Several groups have studied predictors of survival after surgery.[1–5] Two population-based studies have provided data on follow-up since birth, but without distinguishing between preoperative and postoperative survival.[6 7] Such data might be helpful when deciding timing of treatment and during patient selection. This report is based on a national cohort of all 595 Norwegian children with UVH born alive between 1990 and 2015. The patients are followed from birth to initial surgery, through staged surgery and beyond Fontan completion. The aims of the study were to estimate preoperative versus postoperative survival in different UVH subgroups and to explore potential changes in survival between treatment eras.

MATERIALS AND METHODS

Material

Annual number of births in Norway is approximately 60 000 (population 5 million). Oslo University Hospital, Rikshospitalet, was given

national responsibility for registration and treatment of children with complex congenital heart defects (CHD) in 1990. All children with CHD referred to Rikshospitalet were registered in a dedicated clinical database (BERTE) starting in 1990.[8] All operations were recorded prospectively from 1 January 1990 to 31 December 2020, along with dates of death from the Norwegian population registry.[9] Details on causes of death were obtained from BERTE. Among the 72 patients with hypoplastic left heart syndrome (HLHS) operated from 1990 to 2001, 46 (63.9%) were operated in USA (Children's Hospital of Philadelphia),[10] 9 operated at another Norwegian hospital (Haukeland University Hospital) and 5 operated in England (Royal Brompton Hospital and Birmingham Children's Hospital). All these patients have been included in our analyses. The study (IRB number 19/29027) was approved by the Data Protection Officer at our institution, and individual consent for the study was waived.

Patients were identified by their full name and birth date. CHD diagnoses were based on ICD-9 and 10, and when needed, the van Mierop classification.[11] Norwegian children include patients whose parents were immigrants and had been granted Norwegian citizenship.

Patient and public involvement

Patients and the public were not involved in any way in the design, or conduct, or reporting or dissemination plans of our research.

Definitions

The term 'functionally UVH' describes a spectrum of congenital cardiovascular malformations, in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation and another to the pulmonary circulation.[12] UVH diagnoses were based on echocardiography, MRI, invasive studies or findings during surgery. UVH diagnoses were grouped based on an acknowledged congenital heart surgery nomenclature[13]: double inlet left ventricle, double outlet right ventricle (DORV), unbalanced atrioventricular septal defect (uAVSD),HLHS, pulmonary atresia with intact ventricular septum and tricuspid atresia (TA). Diagnoses including less than 40 patients were merged (miscellaneous; MISC). Online supplemental table 1 shows the largest groups of coexistent heart defects. Hypoplastic aortic arch was defined as an aortic arch diameter less than the number of kilogram body weight in millimetres plus 1 millimetre, in at least one segment from the first and to the last aortic arch artery. UVH ascertainment among patients with DORV was done by combining baseline diagnostic data with the actual procedures done.[14]

Since patients with HLHS represent the largest UVH subgroup and undergo specialised surgical correction, we categorised the patients into HLHS and non-HLHS in some analyses.

The initial operations (online supplemental table 2) included: aorto-pulmonary shunt (AP-shunt), ventriculo-pulmonary shunt,[15] banding of the pulmonary artery, bidirectional cavopulmonary connection (BCPC),[16] Norwood 1[17] and others. In 1999–2000, we shifted from lateral tunnel[18] to extracardiac conduit[3 19] during Fontan surgery. In 2002–2003, we shifted from using AP-shunt to ventriculo-pulmonary (Sano) shunt during the classical Norwood 1 operation. Three patients had Bjørk modification of the Fontan procedure.[20]

Home monitoring of nutrition status and oxygen levels among patients with HLHS awaiting BCPC was introduced in 2007. The parents were instructed to register daily body weight and oxygen saturation in a diary and were asked to call a nurse coordinator when significant changes occurred. No patients stayed in hospital continuously between initial surgery and BCPC. Interstage survival was defined from 30 days after stage 1 until the last patient had undergone BCPC. All parents were offered to seek professional psychologic or psychiatric counselling.

Intention to perform surgery

All patients were evaluated by a team of neonatologists, paediatric cardiologists, radiologists, anaesthesiologists and congenital cardiac surgeons. If surgical correction was considered feasible and in agreement with the wishes of the parents, a surgical treatment process was initiated, usually aiming at establishment of Fontan circulation.

We divided reasons for palliative treatment into four categories:

1. General health issues: multiple non-cardiac congenital anomalies, chromosomal defects (trisomy 13, 18) or serious congenital syndromes, and prematurity (gestational age <30 weeks).
2. Surgery not feasible: surgery considered to be associated with too high risk or low probability of a viable result, for example, in presence of major atrioventricular valve insufficiency or limited pulmonary vascular bed.
3. Complications: failure of one or more vital organs (heart, brain, kidney), serious infection (sepsis, pneumonia) or death before surgery could be performed.
4. Parental decision.

Norway has a public healthcare system, which reduced the impact of socioeconomic differences.

Statistical analyses

Kaplan-Meier analyses were used to study survival. The primary end point in the survival analyses was death or heart transplantation (HTX). Survival means 'cumulative survival' if not stated otherwise. The log-rank test (Mantel-Cox) was used to test for differences in survival.

Survival was measured from birth or from time of operation. Right censoring was performed at the end of year 2020 or when reaching a defined age, depending on the context. Competing risk analyses were used when analysing timing of BCPC versus death among patients with HLHS.

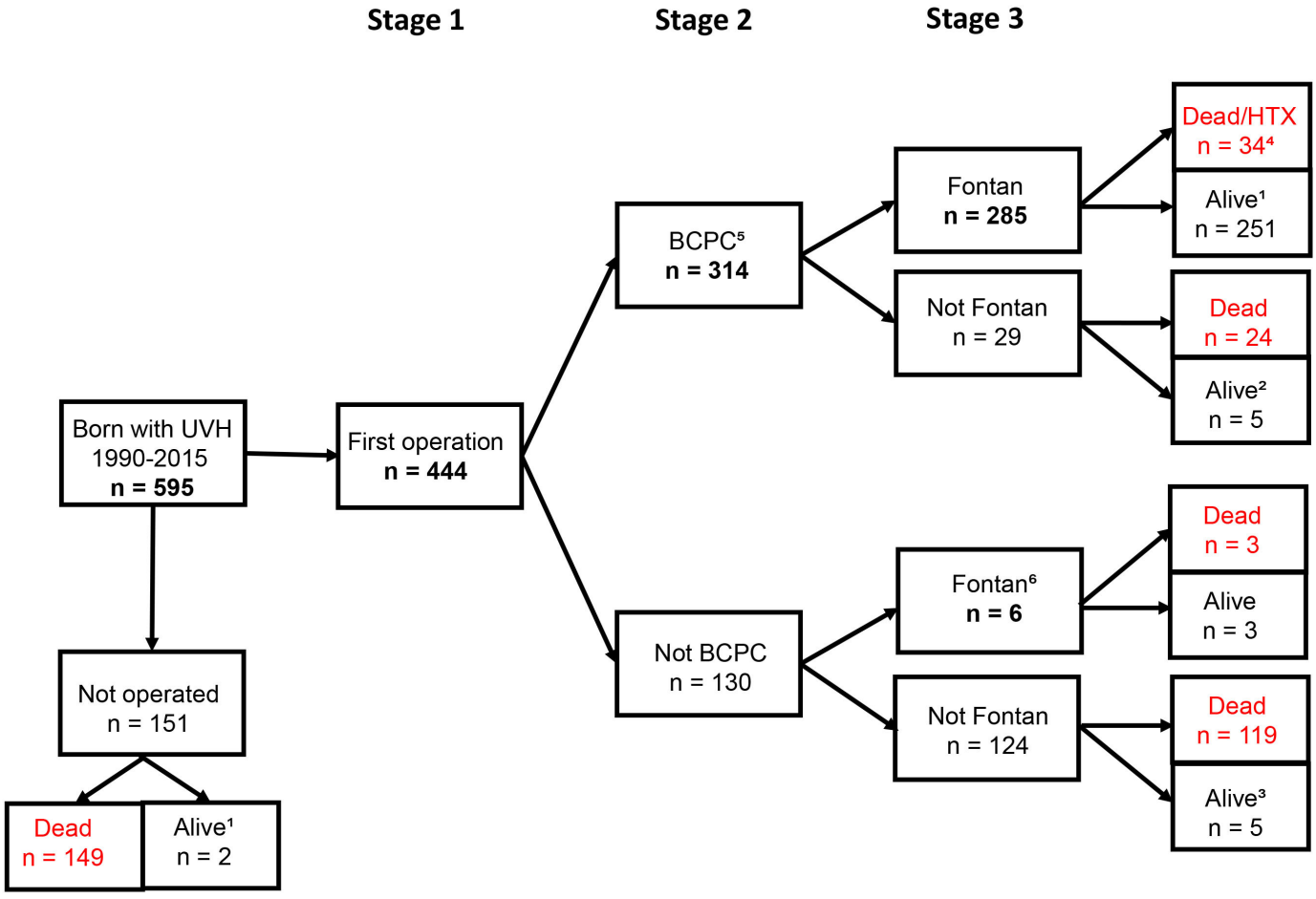


Figure 1 Flowchart showing the number of patients and their outcome during different phases of the study. [1]Both patients pulmonary hypertension. [2]Two patients BCPC as final operation; four two ventricle correction. [3]Five patients with aorto-pulmonary shunt. [4]Thirteen patients heart transplantation (HTX). [5]BCPC (bidirectional cavopulmonar connection) as first operation in 48 patients. [6]Fontan as first operation in 6 patients.

Because of the change of treatment strategies, we dichotomised the data into patients born in 1990–2002 and in 2003–2015.

The Mann-Whitney test was used when comparing clinical variables among survivors.

Two-sided p values <0.05 were considered significant. The statistical package StatView V.5.0 was used in all analyses except the statistical package R for competing risk analyses.

RESULTS

We identified 595 UVH patients born in 1990–2015. Details on annual birth incidence are shown in online supplemental table 3. A total of 270/595 patients (45.5%) were women (one-sided p=0.013).

Figure 1 shows survival through the surgical stages and figure 2 survival in the different diagnostic categories. A total of 283/328 (86.3%) of all deaths occurred during the first 2 years of living.

Before surgery

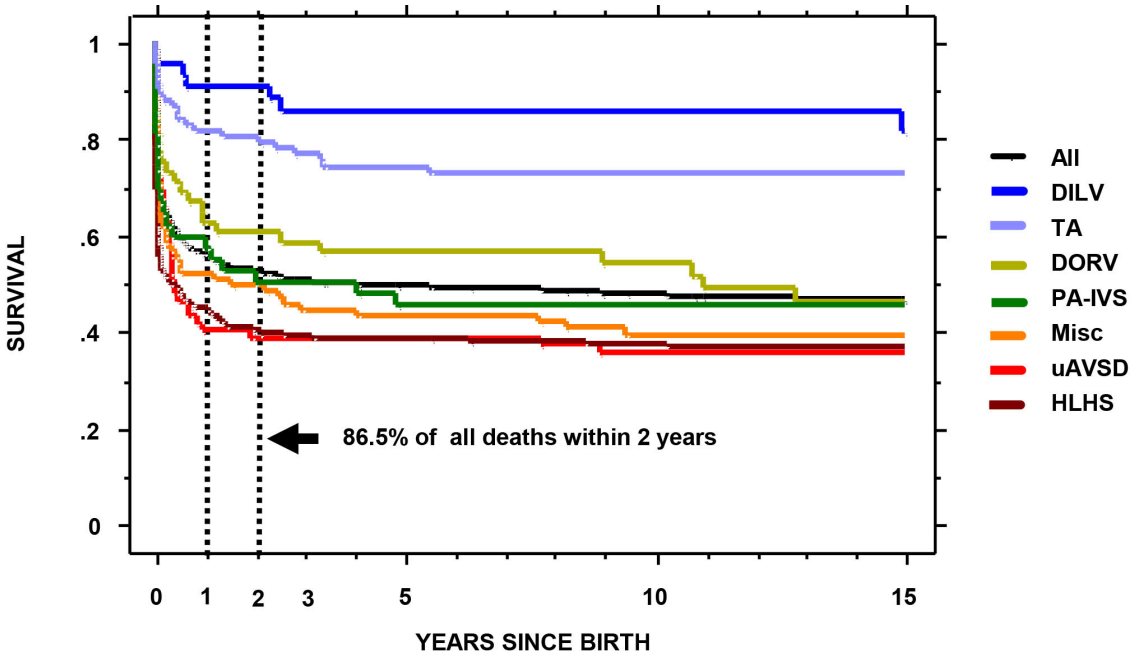
One quarter (151/595; 25.4%) were not operated. Among these, only two patients survived—both having severe pulmonary hypertension by the end of follow-up. A total of 125/149 (83.9%) died within 1 month (figure 3) and 145/149 (97.3%) within 1 year. Median age of death was 7 days, ranging from 3 days in the MISC subgroup (n=19) to 27 days among patients with TA (n=10) (p<0.01).

The proportions of patients not operated varied markedly between the UVH subgroups (table 1). Five of six unoperated patients with Down's syndrome and uAVSD died due to preoperative complications.

The proportions of unoperated patients remained stable during the study, whereas there was an increase in prenatal ultrasound diagnoses (9.9% vs 42.6% among patients born in 1990–2002 vs 2003–2015, p<0.0001). No patients had prenatal treatment.

TGA and aortic arch anomalies were by far the most common coexistent heart defects (21.0% and 12.4%, details not shown). Online supplemental table 1 shows that 79/595 (13.3%) of the patients had serious extracardiac anomalies. The proportions of such anomalies were higher among unoperated than among operated patients (39/151 (25.8%) vs 40/444 (9.0%); p<0.0001).

The reasons to refrain from surgery were that surgery was not considered feasible in 31.1%, preoperative



AT RISK							
• uAVSD	65	26	25	25	25	21	17
• DILV	42	38	38	36	36	30	18
• DORV	48	30	29	28	27	23	15
• HLHS	238	107	95	93	92	70	43
• MISC	79	41	39	35	34	29	20
• PA-IVS	42	25	22	21	19	16	8
• TA	81	66	65	62	60	54	42
• ALL	595	333	313	300	293	243	163

Figure 2 Cumulative 15 years HTX-free survival since birth among subgroups of patients with univentricular heart. DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; HTX, heart transplantation; MISC, miscellaneous; PA-IVS, pulmonary atresia with intact ventricular septum; TA, tricuspid atresia; uAVSD, unbalanced atrioventricular septal defect.

complications in 25.2%, general health issues in 23.2% and parental decision in 20.5%. The decision that surgery was not feasible was the most common cause. A higher proportion of patients with HLHS was not operated due to parental decision compared with patients with non-HLHS (25/81 (30.9%) vs 6/70 (8.6%), p=0.0007). There was no significant difference in the proportions of patients not being operated due to parental decision between patients born in 1990–2002 versus 2003–2015 (19/81 vs 12/70; p=0.33). A decreasing trend among patients with HLHS was borderline significant (19/49 vs 6/32; p=0.056).

Surgery

A total of 444 patients (74.6%) were operated. Variability between the UVH subgroups in postoperative survival was substantial, but preoperative and postoperative survival followed similar patterns (table 1). All patients had a minimum of 5 years of follow-up, and 5-year post-operative HLHS survival was higher among patients born in 2003–2015 than among patients born in 1990–2002, whereas non-HLHS survival did not change significantly (figure 4).

Initial surgery

Median age at initial surgery among patients with HLHS was 6 days, and among patients with non-HLHS was 12 days (p<0.0001). Age at operation was not significantly associated with survival (details not shown). Among patients with HLHS, 30-day survival was much higher among patients with Sano shunt compared with patients with AP-shunt (figure 5). Among the 124 patients who underwent initial surgery but did not have BCPC or TCPC, 119 (96.0%) died, and the five survivors continued to have AP-shunt (figure 1).

Bidirectional cavopulmonary connection (BCPC)

BCPC was established in 314/595 patients (52.8%). Median age at operation among HLHS patients vs. non-HLHS patients was 0.44 years [0.23–0.96] vs 0.80 years [0.22–16.1] (p<0.0001). Forty-eight patients had BCPC as their initial operation. Two patients had BCPC as final operation, and four BCPC patients had subsequent two ventricle repair (figure 1). One year post BCPC survival among HLHS vs non-HLHS patients (Fontan censored) was similar (93.5% vs. 93.7%), and there were no significant associations between age at BCPC and postoperative

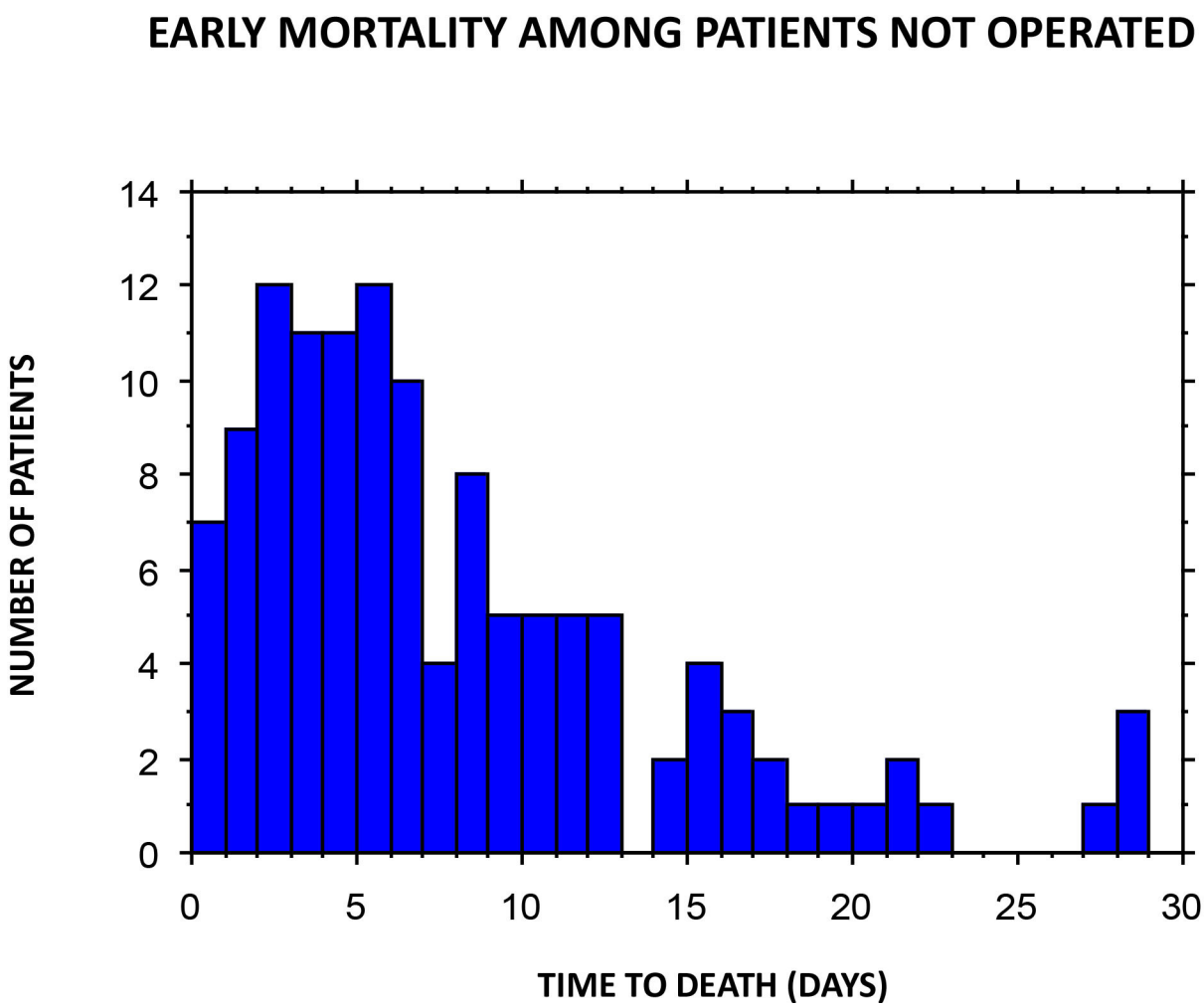


Figure 3 Distribution of time from birth do death within 30 days among patients who were not operated.

survival among HLHS or non-HLHS patients. One-year survival among patients with HLHS surviving 30 days after initial surgery was lower in patients born in 1990–2002 compared with 2003–2015 (76.8% vs 90.1%; p=0.015), and median age at BCPC was higher (0.52 (0.27–1.00) years vs 0.42 (0.23–0.90) years; p=0.0009). Competing risk analyses show that the group with earliest BCPC had lowest interstage mortality (online supplemental figure 1A,B). One-year post-BCPC survival among patients with HLHS born in 1990–2002 vs 2003–2015 was similar (92.9% vs 93.8%).

HLHS survival improved significantly after introduction of home monitoring in 2007. Among patients who survived 30 days after initial surgery, 5 years survival

Table 1 Number of patients not operated, and at initial operation, BCPC and Fontan

UVH subgroups	Number of patients	Operated	BCPC	Fontan	Postoperative survival (5 years)
DILV	42	39 (92.9%)	35 (83.3%)	35 (83.3%)	36 (92.3%)
TA	81	70 (86.4%)	60 (74.1%)	60 (74.1%)	59 (84.3%)
DORV	48	42 (87.5%)	28 (58.3%)	24 (50.0%)	27 (64.3%)
PA-IVS	42	35 (83.3%)	21 (50.0%)	19 (45.2%)	19 (54.3%)
MISC	79	61 (77.2%)	40 (50.6%)	34 (43.0%)	34 (55.7%)
HLHS	238	157 (66.0%)	107 (45.0%)	97 (40.8%)	92 (58.6%)
uAVSD	65	40 (61.5%)	23 (35.4%)	22 (33.8%)	23 (57.5%)
Total	595	444 (74.6%)	314 (52.8%)	291 (48.9%)	308 (69.4%)

DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; MISC, Miscellaneous; PA-IVS, pulmonary atresia with intact ventricular septum; TA, tricuspid atresia; uAVSD, unbalanced atrioventricular septal defect.

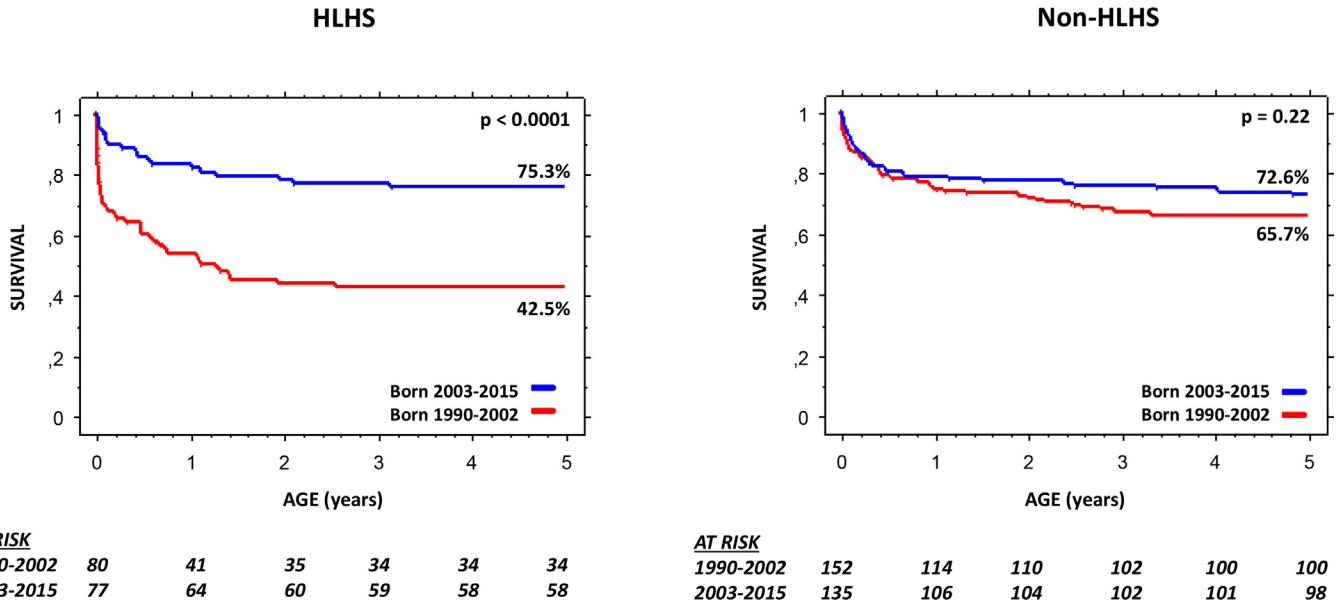


Figure 4 Five years HLHS versus non-HLHS postoperative survival among patients born in 1990–2002 versus 2003–2015. HLHS, hypoplastic left heart syndrome.

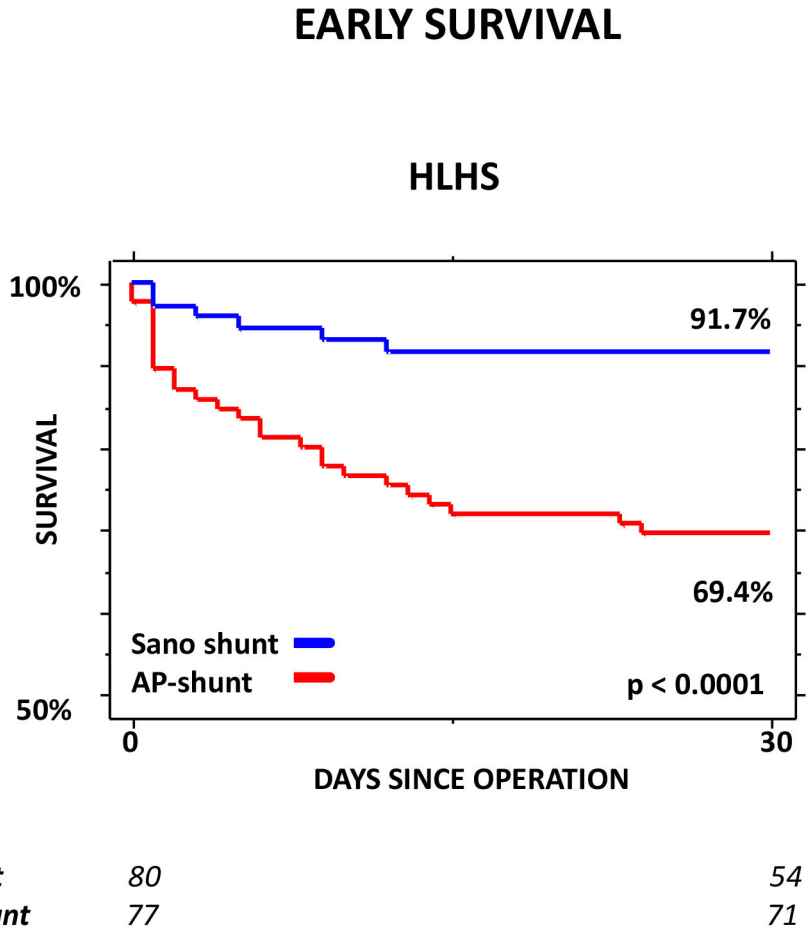


Figure 5 Thirty days postoperative survival among patients with hypoplastic left heart syndrome operated with aorto-pulmonary shunt (AP-shunt) versus Sano shunt. AP, aorto-pulmonary; HLHS, hypoplastic left heart syndrome;

before vs after introduction of home monitoring was 78.2% vs 92.0% (BCPC and Fontan censored; p=0.0135).

Fontan

Fontan surgery was performed in 291/595 patients (48.9%) (HLHS 40.3% vs non-HLHS 52.9%, p=0.003). Median age at operation among patients with HLHS versus patients with non-HLHS was 2.22 years (1.0–13.1) versus 3.19 years (1.1–13.8) (p=0.0008). Six patients had Fontan without BCPC. Survival after Fontan was higher among patients born in 2003–2015 compared with 1990–2002 (online supplemental figure 2). Survival among patients with extracardiac conduit versus lateral tunnel was similar (details not shown). Thirty-four Fontan patients died or had HTX (n=13). Median age at HTX was 16.2 years (7.7–23.9). By the end of follow-up, 10/13 (76.9%) of the HTX patients were still alive (median age 21.4 years (14.4–29.1)). A total of 6/13 were transplanted due to heart failure, 4/13 due to multiorgan failure and 3/13 due to pulmonary vascular dysfunction

Surgical or catheter-assisted interstage procedures

The number of interstage catheter-assisted procedures between initial surgery and BCPC increased from 7/232 (0.9%) to 40/212 (18.9%) (p<0.0001) among patients born in 1990–2002 versus 2003–2015, and the use of such procedures between BCPC and Fontan increased from 8/148 (5.4%) to 25/174 (14.4%) (p=0.008). In contrast, the number of interstage surgical procedures between initial surgery and BCPC and between BCPC and Fontan remained stable around 3–5% (details not shown).

DISCUSSION

This study explores survival in an unselected, nationwide cohort of patients with UVH after birth and through consecutive stages of surgical palliation. It was decided to refrain from surgery in one-fourth of the patients, and survival among these was extremely low. Long-term outcome was mainly determined already during the first 2 years. Preoperative survival varied considerably among the UVH subgroups and followed a similar pattern postoperatively. Improvements in HLHS survival were associated with introduction of the Sano shunt and earlier BCPC. Fontan circulation was established in less than half of the patients.

Before surgery

Only two of the unoperated patients survived. This reflects that UVH is rarely compatible with life if left untreated. Importantly, the public healthcare system in Norway reduced the impact of socioeconomic differences when deciding whether a patient could be operated or not.

The proportions of surgical cases remained stable. Since the use of prenatal ultrasound increased and the detection rates in fetuses with UVH are high,[21] one might have expected increasing proportions of surgical cases.[22] Increasing termination rates[21] following increasing numbers of prenatal ultrasound diagnoses may explain this finding. The increase in termination rates has been later and slower in Norway than, for example, in Denmark.[6 23] In our study, reduction in the number of live-borns was only borderline significant. In a recent study, termination rates increased, but at the same time, there was a sequential increase in proportions of live-borns who underwent surgery.[24] Differences in selection imposed by terminations may impact the proportions of live-born babies in different UVH subgroups, but the mechanisms are complex.

Reasons to refrain from surgery may depend on surgical judgement and/or parental wishes—influences by ethical, cultural and religious factors. Comfort care is a form of active treatment that may reflect family choices and general societal values, which may be particularly important when deciding how to treat children with genetic and other comorbidities. Improved surgical results may increase parent motivation for pursuing surgery. All these factors may influence overall rates of survival. In our study, there was a trend towards less parental decisions against surgery among patients with HLHS in the late era. Interestingly, in a study where all infants with severe CHD were operated regardless of comorbidities, mortality was similar to a study applying a more conservative strategy.[23 25]

Two large previous studies from Denmark and Texas have reported on survival among patients with UVH followed all the way since birth.[6 7] Survival in these studies in comparable eras was similar to ours, although details on preoperative and postoperative survival were not provided. Interestingly, the proportions of patients with serious non-cardiac extracardiac birth defects—which are strongly associated with mortality,[6 7] were similar in these two studies and in our study.

Surgery

Despite considerable variability in postoperative survival among the UVH subgroups, high and low preoperative and postoperative survival were closely correlated. HLHS survival improved considerably during the study period. In contrast, there were no significant changes in the other UVH subgroups—including some groups with a poor prognosis similar to HLHS. This is surprising, since operative and postoperative care might improve as time went by. Moreover, enhanced and more frequent prenatal diagnostics might facilitate surgical planning.[22] However, in a recent study, no significant association between prenatal versus postnatal UVH diagnosis and mortality was found.[26] Our data on postoperative survival are in general agreement with a previous study using a similar UVH subclassification,[27] However, the studies are not directly comparable due to differences in patient selection.

Early HLHS survival was high among patients born in 2003–2015 compared with the 1990–2002. This may in part be explained by the shift from AP-shunt to Sano

shunt during Norwood 1 in 2002–2003. Post Sano survival in our study was similar to previous findings.[1]

Patients with HLHS in the late cohort had earlier BCPC than patients in early cohort, and better interstage survival. Since mortality among patients with HLHS is high, earlier BCPC through better identification of patients at high risk may have prevented death.[28] In our study, introduction of home monitoring of nutrition status and oxygen levels in 2007 was associated with improved survival.[29] It is also conceivable that improved outcomes after initial surgery may have facilitated earlier BCPC. BCPC is currently usually performed at 4–6 months in patients with HLHS similar to our patients,[30] whereas most Patients with non-HLHS are operated at a higher age.[3] Favourable results have been achieved even before 3 months in selected groups with HLHS not being discharged from hospital before BCPC.[30 31]

Less than half of all UVH patients survived until Fontan completion. Previous studies have reported even lower numbers.[6] Accordingly, Fontan patients represent a highly selected UVH subgroup. Post Fontan survival was high, particularly among patients born in the 2003–2015 cohort. Our data are in line with reports from other contemporary Fontan cohorts.[5 32]

Surgical and catheter-assisted interstage procedures

As expected, there was an increase in the number of patients who underwent catheter-assisted procedures. Catheter-assisted and hybrid interventions may reduce the surgical burden and facilitate timing of treatment,[33] and our findings probably reflect recent developments.[34] Importantly, there was no increase in catheterisation facilities at our institution during the study period.

The era effect

During the 30 years study period, there have been a substantial changes in diagnostics and therapeutic methods. In addition to, for example, MRI and echocardiographic diagnostic facilities and advances in perioperative and ICU care, there has been a development in surgical techniques that explain the improved outcomes in our and previous studies. The Sano shunt in HLHS surgery and the extracardiac conduit[32] introduced at our institution around year 2000 probably had an important impact on the results, contributing to the 'era effect' reflected in our study. Interestingly, improved outcomes were mainly seen in the HLHS group, suggesting a potential for further improvements even in other UVH subgroups.

Limitations

UVH defects comprise a complex entity, and a 'correct' morphological diagnosis can be difficult to obtain and may ultimately depend on individual judgement. In the present study, several cardiologists and surgeons were involved during the 26 years inclusion period, and we have not been able to run a complete validity check on all the given diagnoses.

We applied the most commonly used standardised surgical UVH classification scheme[12] similar to previous studies. Still, the diagnostic subgroups are heterogenous, as indicated by the number of coexistent defects in each UVH subgroup.

Reasons to refrain from surgery depend on both surgical and parental judgement, which may be influenced by, for example, ethical, cultural and religious factors. The results from our study reflect the situation in Norway during a defined time period and can not be generalised.

As in all observational studies demonstrating associations, causations can not be implied. Substantial developments, for example, in diagnostics, surgical methods and intensive care during the long inclusion period probably are the main reasons for improvements in outcome.

There was a trend towards lower UVH birth rates during the last part of the study, but we do not know the number of pregnancy terminations (deadline in Norway is 18 weeks).

Conclusions

In this unselected, nationwide cohort of patients with UVH from Norway followed since birth, surgery was refrained in one quarter of the patients, and survival without operation was extremely low. General health issues and preoperative complications accounted for half of the reasons to refrain from surgery, and parental decision and surgical considerations accounted for the rest. HLHS survival improved markedly during the long study period, whereas there were small changes in non-HLHS survival. Preoperative and postoperative survival followed similar patterns among the UVH subgroups.

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Contributors GE, JA, GW, HH and HLL participated in the planning of the study. GE, KL, HH and HLL performed the statistical analyses. GE, GW, GD, SB, TNH, KJ, ES and HLL participated in collection of the data. GE, KL, JA, GW, HH, GD, OG, ET, MEE and HLL participated in the reporting of the work. GE is responsible for the overall content as the guarantor. The guarantor accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

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Patient and public involvement Patients and/or the public were not involved in the design, or conduct, or reporting, or dissemination plans of this research.

Patient consent for publication Not applicable.

Ethics approval The study was approved by the Data Protection Officer at our institution (IRB number 19/29027). As stated above, the study was approved by the Data Protection Officer at our institution (IRB number 19/29027), and individual consent for the study was waived.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available upon reasonable request. Individual participant data after deidentification underlying the results reported in this paper will be shared on reasonable request until 31 December 2029 (eg, through Dryad URL).

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