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Research Article

Mortality and Survival after Norwood Procedure Comparison between Shunt Type in Patients with Hypoplastic Left Heart Syndrome or Its Variants: A Systematic Review and Meta-Analysis Study

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Background. In the Norwood procedure, a conduit is performed either from the subclavian artery to the pulmonary artery, Blalock–Taussig shunt (mBTs), or from the right ventricle to the pulmonary artery (RV-PA shunt). There are some concerns regarding the two shunts and which one is better according to morbidity and mortality in patients with hypoplastic left heart syndrome or its variants. *Methods*. We systematically searched PubMed, Web of Science, Scopus, Embase, and Cochrane Library databases from inception to 04/June/2021 to collect articles reporting a comparison of RV-PA shunt and mBTs. *Results*. Our meta-analysis showed that the mortality rate after 6 months, 1, 2, 3, 4, 5, and 6 years for the mBTS group was 16.3%, 28.6%, 34.8%, 42.4%, 44.6%, 45.1%, and 39.6%, respectively, and for the RV-PAS, 14.8%, 26.6%, 31%, 40.1%, 36.1%, 37.5%, and 34.0%, respectively. The mortality rate was significantly higher in the mBTs group at 1 and 2 years; otherwise, there is no significance differences. Overall complications rate was higher in the mBTs group than in the RV-PAs group (17.8% vs. 8.5%). In contrast, the rate of cardiac complications was higher in the RV-PAS group. *Conclusions*. The RV-PA shunt had lower mortality and overall complications rate than mBT shunt at the short-term outcome within the first two years, but at the long term, there was no difference between the two shunts. On the other hand, the mBT shunt had a lower incidence of cardiac complications at the early stage after the operations. However, some studies are poor due to the difficulties in conducting original research in this field. Therefore, we recommend conducting systematic reviews and original studies to compare these and other therapeutic procedures for these patients.

1. Introduction

Hypoplastic left heart syndrome (HLHS) and its variants are rare congenital heart defects. Children born with a hypoplastic or absent left ventricle may affect the aorta, aortic valve, or mitral valve. HLHS is fatal without early intervention. There are many possible interventions used currently: either a three-stage reconstruction surgery or a heart transplant. Surgical palliation stage 1 was first described by Norwood et al. in 1983 [1].

In the Norwood procedure, the right ventricle is relied on to pump blood to the body and the lungs. To reduce the deoxygenated blood reaching the right ventricle and to make it pass directly to the lungs, a conduit is performed either from the subclavian artery to the pulmonary artery, Blalock–Taussig Shunt (mBTs), or

from the right ventricle to the pulmonary artery (RV-PA shunt).

The Norwood procedure using the mBT shunt appeared as a good first procedure to save these patients, but it did not improve the survival much, so the surgeons looked for other better procedures such as RV-PA shunt, and many primary studies were conducted comparing the two procedures to find which shunt is better to survival and morbidity. Recently, a hybrid procedure appeared that reduced surgical interventions for HLHS patients in the first stage instead of Norwood, but studies and different surgical experiences between different centers are still conflicting about its importance.

There are still some concerns regarding the two shunts and which one is better. Randomized controlled trials and cohort studies show that RV-PA conduit improves transplant-free survival, but morbidity did not affect the outcome between two shunts [2, 3]. A meta-analysis suggested an early survival advantage of RV-PA conduit over the mBTs shunt [4]. In contrast, other studies demonstrated that there is no significant difference between the two shunts [5, 6] and other studies suggest that the mBTs shunt is better [7].

Accordingly, we decided to conduct a systematic review and meta-analysis to try to answer this question comprehensively according to the medical literature.

2. Materials and Methods

We conducted our systematic review and meta-analysis following to recommendations of Preferred Reporting Items for Systematic Reviews and Meta-Analyses 2020 (PRISMA 2020) [8].

2.1. Eligibility Criteria. We include in this systematic review full-text articles reporting a comparison of the RV-PA shunt and mBT shunt in patients with hypoplastic left heart syndrome or its variants who underwent Norwood procedure. We exclude any study that did not mention primary outcome data for both groups. Any study that did not mention mortality, survival, or transplantation was also excluded. The exclusion criteria also contain review articles, case reports, and case series of less than 10 patients. Book chapters, abstract conferences, articles with no full text or missing information, and studies published in languages other than English or Arabic were also excluded.

2.2. Information Sources and Search Strategy. We systematically searched PubMed, Web of Science, Scopus, Embase, and Cochrane Library databases from inception to 04/June/2021 to collect articles reporting a comparison of right ventricle to pulmonary artery conduit and modified Blalock–Taussig shunt. The search terms of our search strategy were showed in Appendix A, and the search strategy was modified to fit each database. No restrictions were applied regarding the date of publication. The reference sections of the included full-text articles were manually evaluated to find more studies eligible for inclusion.

2.3. Selection Process. The title/abstract and full-text screening processes were conducted by two authors separately, and disagreements were resolved by a third author.

2.4. Data Collection Process. Data were extracted from the eligible studies including first author's name, year of publication, journal, country, the intervention and control procedures, number of participants in mBTS shunt/Sano shunt groups, gestational age (weeks), birth weight (kg), surgery stage 1 age (days), sex, ascending aorta diameter (mm), surgery weight (kg) at stage 1, aortic atresia, mitral atresia, aortic stenosis, mitral stenosis, noncardiac anomalies, genetic syndrome, cardiopulmonary bypass time (minutes), aortic cross clamp (minutes), and primary outcome that contain mortality or transplantation, and transplant free survival. Secondary outcomes were reoperation, post-Norwood complications, pacemaker placed, protein-losing enteropathy, and adjudicated cause of death. Data extraction process was performed by one author separately and reviewed by another.

2.5. Quality Assessment of the Studies. For each eligible study, the quality assessment was performed by one investigator and reviewed by another. We assessed the risk of bias in including studies based on Cochrane Collaboration Handbook [9] for RCT studies and the criteria of the Newcastle–Ottawa scale [10] for non-RCT studies. Conflicts are resolved through discussion among authors or the involvement of a third author if necessary.

2.6. Effect Measures and Synthesis Methods. Our primary outcomes were analyzed using the random effect model because of the high level of heterogeneity and with the risk ratio (RR) used as the summary statistic in the forest plots. In our analysis, we used the I^2 statistics to estimate the percentage of total variation in each outcome, owing to heterogeneity rather than chance, with values > 50% considered as substantial heterogeneity. Sensitivity analysis was performed. Publication bias was assessed by funnel plots; any extremity result was excluded from the analysis. All statistical analyses were performed using MetaXL software tool. We could not do meta-analysis for our secondary outcomes because they were reported from one or two studies. We calculated the mortality percentage at 6 months, one, two, three, four, five, and six years by aggregating the number of death events at each time point reported by the included studies divided by the total number of children reported by the included studies.

3. Results

3.1. Identification and Selection of Reviews. We identified 70 articles accepted in our review, after searching in four databases, doing a manual search, screening title/abstract and full text, and applying our criteria on full-text articles. Thirty-four studies have overlap between patients with other studies, so 36 studies were selected for statistical synthesis

including two RCTs and 30 retrospective and four prospective cohort studies. Our PRISMA flow diagram is shown in Figure 1. The main characteristics of the accepted studies are shown in Table 1.

3.2. Characteristic and Quality of the Included Studies. A total of 6183 patients (3144 (50.8%) with RV-PA shunt vs. 3039 (49.2%) with mBTs) were included in all studies. Baseline statistics were between the RV-PA shunt and mBTs groups including gender (RV-PA shunt 890/1431 (60%) male vs. mBTS 749/1180 (59%) male), gestational age (weeks) (RV-PA shunt; mean = 38.68 and SD = 1.48 and $N = 534$ vs. mBTs; mean = 38.95 and SD = 1.42 and $N = 616$), and mean birth weight (kg) (RV-PA shunt; mean = 3.16 and SD = 0.38 and $N = 1160$ vs. mBTs; mean = 3.23 and SD = 0.40 and $N = 1254$).

The other surgical characteristics of the patients were surgery stage 1 age (days) (RV-PA shunt; mean = 7.52 and SD = 3.80 and $N = 1262$ vs. mBTs; mean = 7.26 and SD = 4.21 and $N = 1270$), ascending aorta diameter (mm) (RV-PA shunt; mean = 3.10 and SD = 1.34 and $N = 962$ vs. mBTs; mean = 3.89 and SD = 1.64 and $N = 875$), and cardiopulmonary bypass time (minutes) at stage 1 (RV-PA shunt; mean = 138.46684 and SD = 36.374885 and $N = 582$ vs. mBTs; mean = 132.67 and SD = 40.04 and $N = 497$) Table 2.

The patient's cardiac abnormalities were as follows: hypoplastic left heart syndrome (RV-PA shunt; 1057/1121 (94.3%) vs. mBTs; 1085/1230 (88.2%)), other single ventricle abnormalities (RV-PA shunt; 63/1079 (5.8% vs. mBTs; 146/1160 (12.6%)), and genetic syndrome (RV-PA shunt; 23/388 (5.9%) vs. mBTs; 33/398 (8.3%)). These results are only for studies that mentioned these characteristics in Table 3.

A total of 36 studies were assessed as described above. The only two randomized controlled trials included in our meta-analysis (Single Ventricle Trial and Frommelt et al 2007 study) had a high risk of bias, as selection bias, performance bias, and detection bias were high due to the difficulty of randomization and blinding. Twenty-one of the 29 observational studies we included were considered to be at high quality. In contrast, the remaining eight observational studies were judged to be at poor quality because these studies did not report a comparison of surgery stage 1 age, birth weight, or gestational age between the two groups (Table 4).

3.3. Primary Outcome Results

3.3.1. Mortality. Early mortality is defined as the mortality post operation or in the first 30 days after stage 1 patients (S1P) or prior to hospital discharge. Early mortality was higher in the mBTs group than the RV-PAS group (20.9% from 1410 vs. 13.2% from 1620), (RR, 0.62; 95% CI, 0.50 to 0.75; $p = 0.27$). Our pooled estimate showed mild heterogeneity ($I^2 = 28\%$).

Mortality by 6 months, 1, 2, 3, 4, 5, and 6 years for the mBTs group is 16.3%, 28.6%, 34.8%, 42.4%, 44.6%, 45.1%, and 39.6%, respectively, and for the RV-PAs 14.8%, 26.6%, 31%, 40.1%, 36.1%, 37.5%, and 34.0%, respectively. The difference in survival between the two shunt groups is significantly different at 1 and 2 years; otherwise, there is no significance difference.

All meta-analysis forest plots for this outcome are shown in Figure 2.

3.3.2. Interstage Mortality. The mortality rate after discharge from the hospital after S1P and before stage 2 patients (S2P) was higher in the mBTs group (24.8% from 1264 vs. 16.8% from 1202) (RR, 0.70; 95% CI, 0.58 to 0.83; $p = 0.22$), and the pooled estimate showed mild heterogeneity $I^2 = 19\%$. There was no significant difference between the two groups mortality after S2P and before stage 3 patients (S3P) (7.54%, 6.79%) in the mBTs and RV-PAs cohort, respectively, (RR, 0.93; 95% CI, 0.53 to 1.62; $p = 0.05$) and there was a moderate heterogeneity $I^2 = 48\%$. All meta-analysis forest plots for this outcome are shown in Figure 3.

3.3.3. Duration of Hospital and ICU Stay. There was no significant difference at stage1P (hospital: SMD = 0.08, $P \leq 0.001$, 95% CI: (−0.26–-0.41)) (mean hospital days RV-PAs = 32.5, SD = 18.8 vs. mean hospital days mBTs = 29.4, SD = 16.7); ICU: SMD = 0.22, $P = 0.01$, 95% CI: (−0.27, 0.71) (mean ICU days RV-PAs = 18.85, SD = 15.3 vs. mean ICU days mBTs = 17.6, SD = 12.95), S2P (hospital: SMD = 0.12, $P = 0.80$, 95% CI: −0.09, 0.33 (mean hospital days RV-PAs = 24.4, SD = 4.7 vs. mean hospital days mBTs = 28.9, SD = 26.3); ICU: SMD = 0.29, $P = 0.04$, 95% CI: −0.52, 1.09) (mean ICU days RV-PAs = 11.6, SD = 6.6 vs. mean ICU days mBTs = 11.3, SD = 6.9), and higher in the RV-PAs group at S3P (ICU: SMD = 0.36, $P = 0.83$, 95% CI: −0.15, 0.57) (mean ICU days RV-PAs = 12.5, SD = 7.1 vs. mean ICU days mBTs = 11.4, SD = 6.9) (Figure 3).

3.4. Secondary Outcome Results (Complications and Adjudicated Causes of Death). Overall complications rate was higher in the mBTs group than in the RV-PAs group (17.8% Vs. 8.5%). The RV-PAs group had less gastrointestinal complications than the mBTs group (3.9% vs. 8.47%). In contrast, the rate of cardiac complications, respiratory complications, neurological complications, infectious complications, and renal complications were higher in the RV-PAs group than in the mBTs group as shown in Table 2. As for the other complications, there was no significant difference between the two groups. Furthermore, pacemakers placing rates were slightly higher in the mBTs group (2.5% vs. 2.90%), whereas RV-PAs patients were more likely to develop the protein-losing enteropathy (3.3% vs. 1.59%). Only one study reported the adjudicated causes of death. Mortality from pulmonary, neurological, gastroenterological, and infectious causes was higher in the RV-PAs group, whereas participants in the mBTs group were more likely to die of cardiac or complex/multisystem causes. On the other hand, the death rate from unknown causes was equal between the two groups (1.8% vs. 1.81%), while there was no study reported the death of organ failure cause. The details of

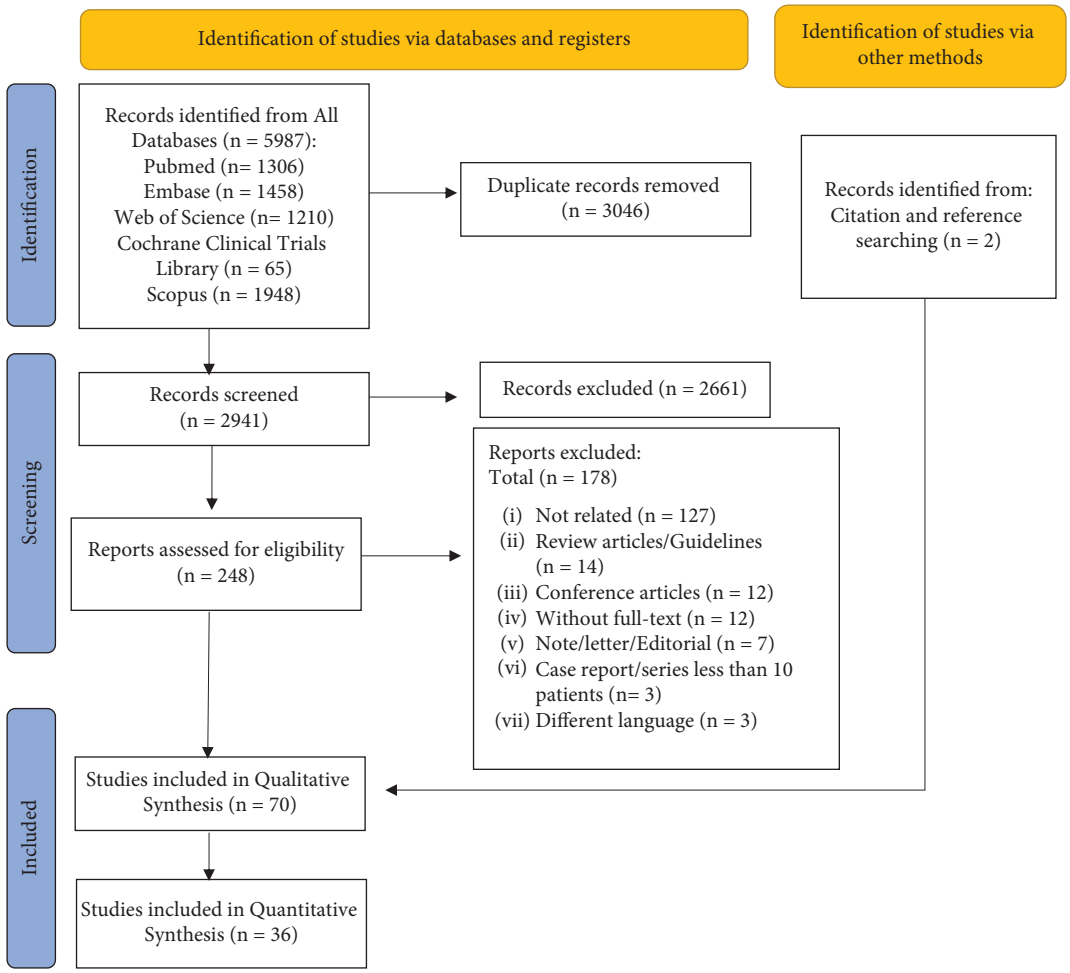

Figure 1: Prism flow diagram of the study.

the complications and the adjudicated causes of death data are shown in Table 5.

4. Discussion

The Norwood procedure is the first of the series of three operations which involve performing two different systemic to pulmonary shunts to ensure pulmonary blood flow in children with HLHS, either mBT shunt or RV-PA shunt. The advantage of either shunt over the other has long been a matter of debate, especially with regard to the long-term outcomes. The results of our meta-analysis indicate the advantage of using the RV-PA shunt to reduce early mortality after S1P and interstage mortality after S1P and before S2P. As for interstage mortality after S2P and before S3P, there was no preference for either of the two shunts over the other. Our meta-analysis provides a look at the mortality after 6 months, 1, 2, 3, 4, 5, and 6 years, in which there was significant difference at 1 and 2 years; otherwise, there is no significance difference.

Furthermore, mBT shunt was accused of causing higher overall complications rate, and this was confirmed by the results of our study, with varying incidence of these complications in both groups; while the patients who received mBT shunt are more likely to develop gastrointestinal complications, and the patients who received RV-PA shunt are more likely to develop cardiac, respiratory, neurological, infectious, and renal complications, and maybe this is what made some centers use the mBT shunt until now. Because of the small number of studies that reported this outcome, the differences in complications between the two groups cannot be explained and can be attributed to the difference in medical centers and surgical experience in performing these two procedures.

As for the causes of death, our review found only one paper that reported the various causes of death in both groups, [14] where children with mBT shunt were more likely to die of cardiac or complex/multisystem causes, while children in RV-PA shunt group were more likely to die from other causes.

The RV-PA shunt is associated with a lower mortality rate in the first 30 days after S1P or prior to hospital discharge. [4] In addition, the results of the single ventricle trial, which was conducted on 549 children, have shown that the transplantation-free survival at one year was better with the RV-PA shunt. In contrast, the current evidence shows similar results between the two groups after one year of age [3, 7]. Although studies have indicated that patients of the

Table 1: Studies' characteristics.

Study ID	Last name, year	Study design	Database/Unit	Country	Study timeline
1	Ono et al., 2021 [11]	Retrospective cohort study	German Heart center Munich	Germany	2001–2019
2	Vitanova et al., 2019 [6]	Retrospective cohort study	German Heart center Munich	Germany	2001–2017
3	Kelly et al., 2019 [12]	Retrospective cohort study	The Royal Children's Hospital	Australia	Era 1: 2004-2008. Era 2: 2008–2016
4	Anton-Martin et al., 2019 [13]	Retrospective cohort study	Phoenix Children's Hospital	USA	2013–2016
5	Newburger et al., 2018 [14]	RCT	Pediatric heart network	USA and Canada	2005–2008
6	Mahle et al., 2018 [5]	RCT	Pediatric heart network	USA and Canada	2005–2008
7	Hall et al., 2017 [15]	Prospective cohort study	Vanderbilt University Medical center	USA	2007–2015
8	Buelow et al., 2018 [16]	Prospective cohort study	National Pediatric Cardiology Quality Improvement Collaborative, Children's Hospital of Wisconsin	USA	2008–2014
9	Wilder et al., 2017 [17]	Prospective cohort study	Congenital Heart Surgeons' Society Data center	USA	2005–2014
10	Ruotsalainena et al., 2017 [18]	Retrospective cohort study	Children's Hospital, University of Helsinki, Helsinki, Finland	Finland	2003–2010
11	Ravishankar et al., 2016 [19]	RCT	Pediatric heart network	USA, Canada	2005–2008
12	Murthy et al., 2016 [20]	RCT	Pediatric heart network	USA, Canada	2006–2013
13	Carlo et al., 2016 [21]	Prospective cohort study	The Pediatric Heart Transplant Study (PHTS) database	USA, UK, Canada	2010–2013
14	Wilder et al., 2015 [22]	Prospective cohort study	Congenital Heart Surgeons' Society Data center	Canada, USA	2005–2014
15	Newburger et al., 2014 [7]	RCT	Pediatric heart network	USA, Canada	2005–2008
16	Burch et al., 2014 [23]	RCT	Pediatric heart network	USA, Canada	2005–2008
17	Gist et al., 2013 [24]	Retrospective cohort study	Children's Hospital Colorado	USA	2003–2011
18	Frommelt et al., 2013 [25]	RCT	Pediatric heart network	USA, Canada	2005–2008
19	Fischbach et al., 2012 [26]	Retrospective cohort study	German Pediatric Heart center	Germany	2002–2009
20	Tweddell et al., 2012 [27]	RCT	Pediatric heart network	USA, Canada	2005–2008
21	Tabbutt et al., 2012 [28]	RCT	Pediatric heart network	USA, Canada	2005–2008
22	Polimenakos et al., 2012 [29]	Retrospective cohort study	Advocate Hope Children's Hospital	USA	2005–2009
23	Photiadis et al., 2012 [30]	Retrospective cohort study	German Pediatric Heart center	Germany	2002–2009
24	Ohye et al., 2012 [31]	RCT	Pediatric heart network	USA, Canada	2005–2008
25	Newburger et al., 2012 [32]	RCT	Pediatric heart network	USA, Canada	2005–2008
26	Murtuza et al., 2012 [33]	Retrospective cohort study	Birmingham Children's Hospital	UK	2002–2010
27	Menon et al., 2012 [34]	Retrospective cohort study	Utah and the intermountain West region	USA	1995–2010
28	Ghanayem et al., 2012 [35]	RCT	Pediatric heart network	USA, Canada	2005–2008

Table 1: Continued.

Study ID	Last name, year	Study design	Database/Unit	Country	Study timeline
29	Rüffer et al., 2011 [36]	Retrospective cohort study	University Heart center Hamburg	Germany	1998–2007
30	Johnson et al 2011 [37]	RCT	Pediatric heart network	USA	2005–2008
31	Fiore et al., 2011 [38]	Retrospective cohort study	Cardinal Glennon Children's Hospital	USA	2000–2007
32	Bautista-Hernandez et al., 2011 [39]	Retrospective cohort study	Children's Hospital Boston	USA	2000–2007
33	Ohye et al., 2010 [3]	RCT	Pediatric heart network	USA, Canada	2005–2008
34	Graham et al., 2010 [40]	Retrospective cohort study	Medical University of South Carolina	USA	2000–2005
35	Ballweg et al., 2010 [41]	Retrospective cohort study	The Children's Hospital of Philadelphia	USA	2002–2005
36	Rüffer et al., 2009 [42]	Retrospective cohort study	University Hospital Hamburg-Eppendorf	Germany	1997–2006
37	Pruetz et al., 2009 [43]	Retrospective cohort study	Children's Hospital Los Angeles	USA	2000–2005
38	Fabricius et al., 2009 [44]	Retrospective cohort study	Birmingham Children's Hospital	UK	1992–2007
39	Al-Akhfash et al., 2009 [45]	Retrospective cohort study	King Abdulaziz Ccardiac center	Saudi Arabia	2001–2007
40	Scheurer et al., 2008 [46]	Retrospective cohort study	Boston Children's Hospital	USA	2002-2003
41	Caspi et al., 2008 [47]	Retrospective cohort study	Louisiana State University and Children's Hospital	USA	2000–2007
42	Atallah et al., 2008 [48]	Prospective cohort study	The Stollery Children's Hospital	Canada	1996–2005
43	Silva et al., 2007 [49]	Retrospective cohort study	Hospital Beneficência Portuguesa de São Paulo	Brazil	1999–2006
44	Silva et al., 2007 [50]	Retrospective cohort study	Hospital Beneficência Portuguesa de São Paulo	Brazil	1999–2006
45	Lai et al., 2007 [51]	Retrospective cohort study	Boston Children's Hospital	USA	2002-2003
46	Kussman et al., 2007 [52]	Retrospective cohort study	Boston Children's Hospital	USA	2002–2004
47	Januszewska et al., 2007 [53]	Retrospective cohort study	Jagiellonian University	Poland	1995–2006
48	Graham et al., 2007 [54]	Retrospective cohort study	Medical University of South Carolina	USA	2000–2005
49	Frommelt et al., 2007 [55]	RCT	Children's Hospital of Wisconsin	USA	2004-2005
50	Edwards et al., 2007 [56]	Retrospective cohort study	Birmingham Children's Hospital	UK	2001–2003
51	Filippo et al., 2007 [57]	Retrospective cohort study	Children's Hospital of Pittsburgh	USA	2000–2005

Table 1: Continued.

Study ID	Last name, year	Study design	Database/Unit	Country	Study timeline
52	Ballweg et al., 2007 [58]	Retrospective cohort study	Children's Hospital of Philadelphia	USA	2002–2005
53	Pusca et al., 2006 [59]	Retrospective cohort study	Emory University	USA	1999–2005
54	Petit et al., 2006 [60]	Retrospective cohort study	The Children's Hospital of Philadelphia	USA	2002–2005
55	Griselli et al., 2006 [61]	Retrospective cohort study	Wales Children's Hospital	UK	1992–2004
56	Griselli et al., 2006 [62]	Retrospective cohort study	Wales Children's Hospital	UK	1992–2004
57	Ghanayem et al., 2006 [63]	RCT	Children's Hospital of Wisconsin	USA	2004-2005
58	Cua et al., 2006 [64]	Retrospective cohort study	Boston Children's Hospital	USA	2002-2003
59	Tabbutt et al., 2005 [65]	Retrospective cohort study	Children's Hospital of Philadelphia	USA	2002–2004
60	Januszewska et al., 2005 [66]	Retrospective cohort study	Jagiellonian University	Poland	1997–2004
61	Tanoue et al., 2004 [67]	Retrospective cohort study	Fukuoka Children's Hospital Medical center	Japan	1992–2003
62	Pizarro et al., 2004 [68]	Retrospective cohort study	Nemours Cardiac center	Poland	2000–2003
63	Hughes et al., 2004 [69]	Prospective case series	Royal Children's Hospital	Australia	2000–2003
64	Bradley et al., 2004 [70]	Retrospective cohort study	Medical University of South Carolina	USA	2000–2003
65	Pizarro et al., 2003 [71]	Retrospective cohort study	Nemours Cardiac center Wilmington, the Polish American Hospital in Krakow	USA, Poland	2000–2002
66	Malec et al., 2003 [72]	Retrospective cohort study	Jagiellonian University	Poland	1998–2002
67	Mair et al., 2003 [73]	Retrospective case series	General Hospital Linz	Austria	1999–2002
68	Mahle et al., 2003 [74]	Retrospective cohort study	Emory University School of Medicine	USA	1999–2002
69	Graham et al., 2005 [75]	Retrospective cohort study	Medical University of South Carolina	USA	2000–2005
70	Azakie et al., 2004 [76]	Retrospective cohort study	San Francisco Children's Hospital and Pediatric Cardiac Program	USA	2001–2003

RCT: randomized control trial; USA: United States of America; UK: United Kingdom.

Table 2: Patient's baseline characteristics.

Study's ID	Last name, year	Population			Gestational age (weeks)								
		RV-PA conduit	Modified BT shunt	Total	RV-PA conduit			Modified BT shunt			Together		
					Mean	SD	N	Mean	SD	N	Mean	SD	N
1 2\19\23	Ono et al., 2021	163	159	322	39	1.496	163	39	1.496	159	39	1.496	322
	Kelly et al., 2019	45	88	133	38.1	1.7	45	39.2	1.5	88	NR	NR	NR
3	Era 1	35	22	57	38.3	1.7	35	39.5	1	22	NR	NR	NR
	Era 2	10	66	76	37.7	1.8	10	39.1	1.5	66	NR	NR	NR
4	Anton-Martin et al., 2019	20	14	34	38.66	0.74	20	39	0.74	7	38.6	0.7	34
5\6\11\15\16\18\20\21\24\25\28\30\33	Newburger et al., 2018	274	275	549	NR	NR	NR	NR	NR	NR	NR	NR	NR
7	Hall et al., 2018	58	62	120	38.7	1.11	58	38.7	1.03	62	38.7	1.05	120
8	Buelow et al., 2018	691	466	1157	NR	NR	NR	NR	NR	NR	38.5	1.5	1156
9	Wilder et al., 2017	222	232	454	NR	NR	NR	NR	NR	NR	NR	NR	NR
10	Ruotsalainena et al., 2017	40	23	63	NR	NR	NR	NR	NR	NR	NR	NR	NR
12	Murthy et al., 2016	75	47	122	NR	NR	NR	NR	NR	NR	NR	NR	NR
13	Carlo et al., 2016	111	79	190	NR	NR	NR	NR	NR	NR	NR	NR	NR
14	Wilder et al., 2015	169	169	338	NR	NR	NR	NR	NR	NR	NR	NR	NR
17	Gist et al., 2013	67	33	100	38.3	1.5	67	38.3	1.3	33	NR	NR	NR
22	Polimenakos et al., 2012	65	25	90	NR	NR	NR	NR	NR	NR	NR	NR	NR
26\38\50	B. Murtuza et al., 2012	264	84	348	NR	NR	NR	NR	NR	NR	NR	NR	NR
27	Menon et al., 2012	93	74	167	NR	NR	NR	NR	NR	NR	38	2.5	245
29\36	Rüffer et al., 2011	25	33	58	39	2	25	40	1	33	NR	NR	NR
31	Fiore et al., 2011	15	19	34	40	1.1	15	39	1.6	19	NR	NR	NR
32\40\45\46\58	Bautista-Hernandez et al., 2011	36	82	118	NR	NR	NR	NR	NR	NR	NR	NR	NR
34\48\64\69	Graham et al., 2010	41	35	76	38.3	NR	41	38.2	NR	35	NR	NR	NR
35\52\54\59	Ballweg et al., 2010	62	114	176	NR	NR	NR	NR	NR	NR	NR	NR	NR
37	Pruetz et al., 2009	32	32	64	NR	NR	NR	NR	NR	NR	NR	NR	NR
38	A.M. Fabricius et al., 2009	28	244	272	NR	NR	NR	NR	NR	NR	NR	NR	NR
39	Al-Akhfash et al., 2008	22	6	28	NR	NR	NR	NR	NR	NR	NR	NR	NR
41	Caspi et al., 2008	23	19	42	NR	NR	NR	NR	NR	NR	NR	NR	NR
42	Atallah et al., 2008	32	62	94	39.4	1.4	32	38.8	1.7	62	NR	NR	NR
43\44	Silva, JP et al., 2007	41	37	78	NR	NR	NR	NR	NR	NR	NR	NR	NR
47\60	Januszewska 2007	150	51	201	NR	NR	NR	NR	NR	NR	NR	NR	NR
49\57	Frommelt 2007	9	8	17	NR	NR	NR	NR	NR	NR	NR	NR	NR
51	Filippo 2007	14	19	33	38.5	1	14	38	1.7	19	NR	NR	NR
53\68	Pusca 2006	30	29	59	NR	NR	NR	NR	NR	NR	NR	NR	NR
55\56	Griselli 2006	74	293	367	NR	NR	NR	NR	NR	NR	NR	NR	NR
70	Azakie et al., 2004	13	8	21	NR	NR	NR	NR	NR	NR	NR	NR	NR
61	Tanoue et al., 2004	39	23	62	NR	NR	NR	NR	NR	NR	NR	NR	NR
62\65	Pizarro et al., 2004	50	46	96	38.1	1.7	50	38.4	1.6	46	NR	NR	NR
66	Malec et al., 2003	37	31	68	NR	NR	NR	NR	NR	NR	NR	NR	NR
67	Mair et al., 2003	14	18	32	NR	NR	NR	NR	NR	NR	NR	NR	NR

RV-PA conduit: right ventricle to pulmonary artery conduit; modified BT shunt: modified Blalock–Taussig shunt; NR: not reported.

Table 3: Patient's baseline characteristics (complete).

Study's ID	Last name, year	Birth weight (kg)						Sex					
		RV-PA conduit			Modified BT shunt			RV-PA conduit			Modified BT shunt		
		Mean	SD	N	Mean	SD	N	Mean	SD	N	Mean	SD	N
1\2\19\23	Ono et al., 2021	3.2	0.449	163	3.167	0.524	159	NR	NR	NR	NR	NR	NR
	Kelly et al., 2019	2.98	0.68	45	3.45	0.53	88	NR	NR	NR	NR	NR	NR
3	Era 1	3.09	0.67	35	3.63	0.39	22	NR	NR	NR	NR	NR	NR
	Era 2	2.85	0.68	10	3.44	0.53	66	NR	NR	NR	NR	NR	NR
4	Anton-Martin et al., 2019	NR	NR	NR	NR	NR	NR	11	9	20	5	2	7
5\6\11\15\16\18\20\21\24\25\28\30\33	Newburger et al., 2018	3.1	NR	274	3.1	NR	275	167	107	274	173	102	275
7	Hall et al., 2018	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
8	Buelow et al., 2018	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
9	Wilder et al., 2017	3.15	0.47	222	3.2	0.52	232	145	77	222	147	85	232
10	Ruotsalainena et al., 2017	3.5	0.4	40	3.5	0.4	23	23	17	40	13	10	23
12	Murthy et al., 2016	3.18	0.47	75	3.34	0.59	47	NR	NR	NR	NR	NR	NR
13	Carlo et al., 2016	NR	NR	NR	NR	NR	NR	58	53	111	48	31	79
14	Wilder et al., 2015	3.2	0.5	169	3.2	0.5	169	105	64	169	113	56	169
17	Gist et al., 2013	NR	NR	NR	NR	NR	NR	37	30	67	20	13	33
22	Polimenakos et al., 2012	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
26\38\50	B. Murtuza et al., 2012	NR	NR	NR	NR	NR	NR	163	101	264	57	27	84
27	Menon et al., 2012	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
29\36	Rüffer et al., 2011	3.258	0.445	25	3.371	0.483	33	14	11	25	23	10	33
31	Fiore et al., 2011	3.3	0.5	15	3.3	0.5	19	NR	NR	NR	NR	NR	NR
32\40\45\46\58	Bautista-Hernandez et al., 2011	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
34\48\64\69	Graham et al., 2010	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
35\52\54\59	Ballweg et al. 2010	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
37	Pruetz et al., 2009	2.96	0.6	32	3.11	0.44	32	22	10	32	18	14	32
38	A.M. Fabricius et al., 2009	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
39	Al-Akhfash et al., 2008	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
41	Caspi et al., 2008	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
42	Atallah et al., 2008	3.4	0.5	32	3.2	0.6	62	21	11	32	33	29	62
43\44	Silva, JP et al., 2007	NR	NR	NR	NR	NR	NR	23	18	41	18	19	37
47\60	Januszewska 2007	NR	NR	NR	NR	NR	NR	25	6	31	14	5	19
49\57	Frommelt 2007	3.1	0.6	9	3.5	0.5	8	5	4	9	6	2	8
51	Filippo 2007	3.3	0.5	14	3.1	0.4	19	11	3	14	15	4	19
53\68	Pusca 2006	NR	NR	NR	NR	NR	NR	22	8	30	18	11	29
55\56	Griselli 2006	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
70	Azakie et al., 2004	NR	NR	NR	NR	NR	NR	9	4	13	6	2	8
61	Tanoue et al., 2004	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
62\65	Pizarro et al., 2004	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR
66	Malec et al., 2003	NR	NR	NR	NR	NR	NR	29	8	37	22	9	31
67	Mair et al., 2003	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR	NR

RV-PA conduit: right ventricle to pulmonary artery conduit; modified BT shunt: modified Blalock–Taussig shunt; NR: not reported.

Table 4: Quality assessment of selected studies using the Newcastle–Ottawa Scale.

ID	Study	S1	S2	S3	S4	C	O1	O2	O3	Total	Quality score
1\2\19\23	One et al., 2021	*a	*a	*a	*a	**	*b	*a	*b	9/9	High quality
3	Kelly et al., 2020	*a	*a	*a	*a	*	*b	*a	*b	8/9	High quality
4	Anton-Martin et al., 2019	*a	*a	*a	*a	**	d	*a	*b	8/9	High quality
7	Hall et al., 2017	*a	*a	*a	*a	**	c	*a	*b	8/9	High quality
8	Buelow et al., 2018	*a	*a	*a	*a	-	d	*a	*b	6/9	Poor quality
9	Wilder et al., 2016	*a	*a	*a	*a	-	d	*a	*b	6/9	Poor quality
10	Ruotsalainen et al., 2017	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
12	Murthy et al., 2015	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
13	Carlo et al.. 2016	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
14	Wilder et al., 2015	*a	*a	*a	*a	*	c	*a	*b	7/9	High quality
17	Gist et al., 2013	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
22	Polimenakos et al., 2012	*a	*a	*a	*a	*	*a	*a	*b	8/9	High quality
26\38\50	Murtuza et al., 2012	*a	*a	*a	*a	-	*a	*a	*b	7/9	Poor quality
27	Shaji et al., 2012	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
29 \ 36	Rüffer et al., 2011	b	*a	*a	*a	**	*a	*a	*b	9/9	High quality
31	Fiore et al., 2011	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
32\40\45\46\58	Bautista-Hernandez et al., 2011	*a	*a	*a	*a	-	*a	*a	*b	7/9	Poor quality
34\48\64\69	Graham et al., 2010	*a	*a	*a	*a	*	*a	*a	*b	8/9	High quality
35\52\54\59	Ballweg et al., 2010	*a	*a	*a	*a	*	*a	*a	*b	8/9	High quality
37	Pruetz et al., 2009	*a	*a	*a	*a	**	*b	b	*b	8/9	High quality
38	Fabricius et al., 2009	b	*a	*a	*a	-	d	*a	*b	5/9	Poor quality
39	Al-Akhfash et al., 2009	*a	*a	*a	*a	*	*a	*a	*b	8/9	High quality
41	Caspi et al., 2008	*a	*a	*a	*a	-	d	b	*b	5/9	Poor quality
42	Atallah et al., 2008	*a	*a	*a	*a	**	*a	*a	*b	9/9	High quality
43\44	Silva et al., 2007	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
47\60	Januszewska et al., 2007	*a	*a	*a	*a	-	*b	*a	*b	7/9	Poor quality
51	Filippo et al., 2007	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
53\68	Pusca 2006	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
55\56	Griselli 2006	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
70	Azakie et al., 2004	*a	*a	*a	*a	-	*a	*a	*a	7/9	Poor quality
61	Tanoue et al., 2004	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
62\65	Pizarro et al., 2004	*a	*a	*a	*	*	*a	b	*b	7/9	High quality
66	Malec et al., 2003	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality
67	Mair et al., 2003	*a	*a	*a	*a	*	*a	b	*b	7/9	High quality

More stars (*) indicate higher quality of study. S1, representativeness of exposed cohort; S2, selection of nonexposed cohort; S3, ascertainment of exposure; S4, demonstration that outcome of interest was not present at the start of study; C1, comparability of cohort on basis of design or analysis; O1, assessment of outcome; O2, follow-up long enough for outcomes to occur; O3, adequacy of follow-up. Adjust for: (1) comparability: (1) surgery stage 1 age (days), (2) birth weight (kg)/gastational age (weeks); (2) follow-up period: at least 1 year for mortality and transplantation; (3) adequacy of follow-up of cohorts: lost subjects <5%. We considered studies as follows: Good quality: 3 or 4 stars in selection domain AND 1 or 2 stars in comparability domain AND 2 or 3 stars in outcome/exposure domain. Fair quality: 2 stars in selection domain AND 1 or 2 stars in comparability domain AND 2 or 3 stars in outcome/exposure domain. Poor quality: 0 or 1 star in selection domain OR 0 stars in comparability domain OR 0 or 1 stars in outcome/exposure domain.

RV-PA shunt group are relatively less likely to develop complications, caution should be taken against shunt failure, especially during the first week [6]. This is in addition to the serious neurological complications associated with this type of shunt, such as seizure, bleeding, and stroke [7, 22]. On the other hand and in the context of constant attempts to develop care for patients with HLHS, the Hybrid procedure has emerged in recent years to be sometimes an alternative to the Norwood procedure [77]. Hybrid procedure shows a good midterm survival in high risk patients [77, 78].

There are several limitations to the current study. These limitations include the lack of high-quality randomized controlled trials due to the nature of the surgical procedure that limits the application of blinding and randomization, there are also some observational studies of poor quality in our review. The heterogeneity that is sometimes present in some pooling operations, also limits this study. In addition, many of the studies included in the review did not discuss many important details for HLHS patients after the procedure such as the diameter of the artery, pulmonary artery

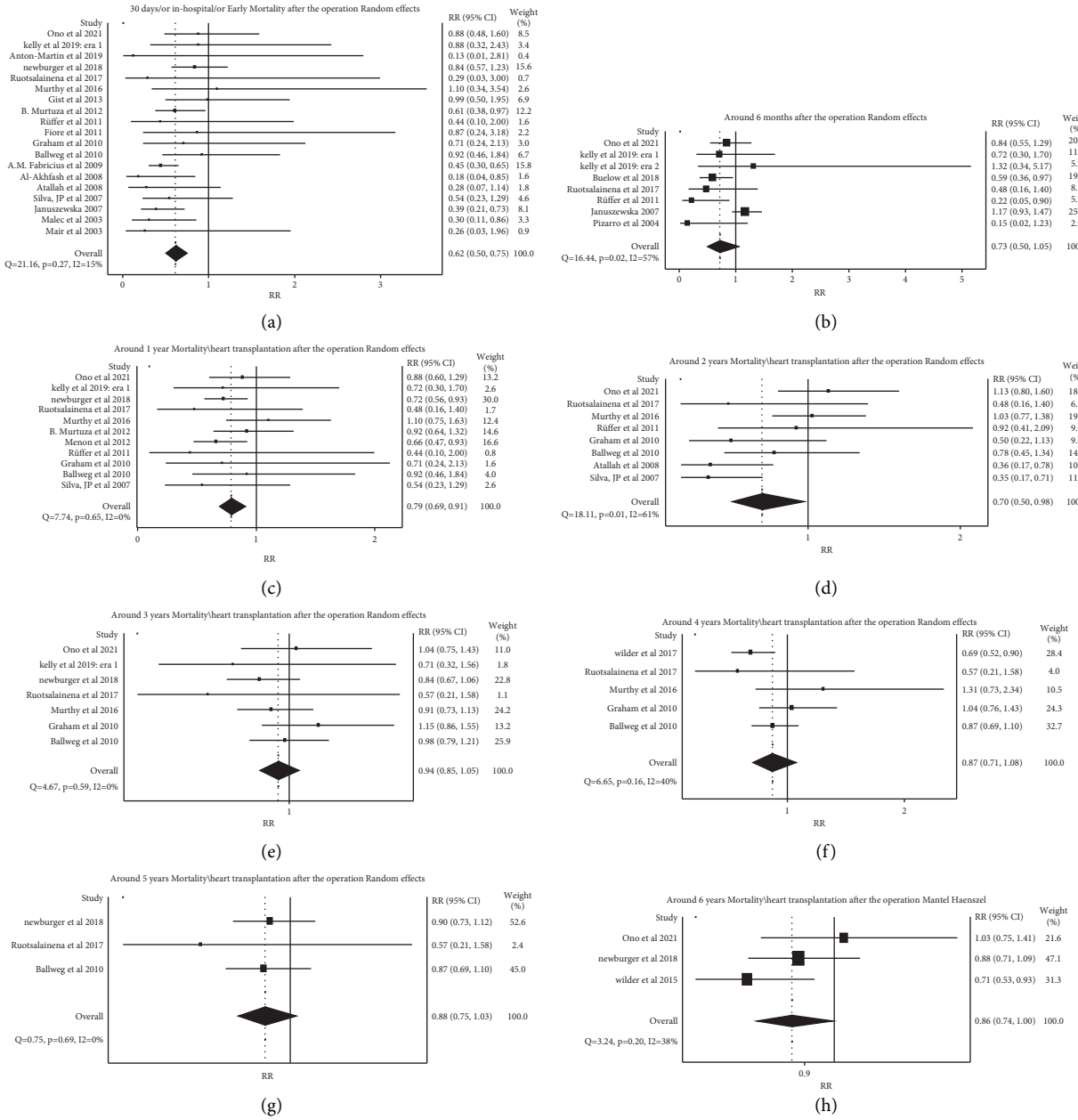

Figure 2: Forest plot of mortality outcomes after 30 days, 6 months, one year, two years, three years, four years, five years, and six years from the operation. Right ventricle to pulmonary artery shunt vs. modified Blalock–Taussig shunt.

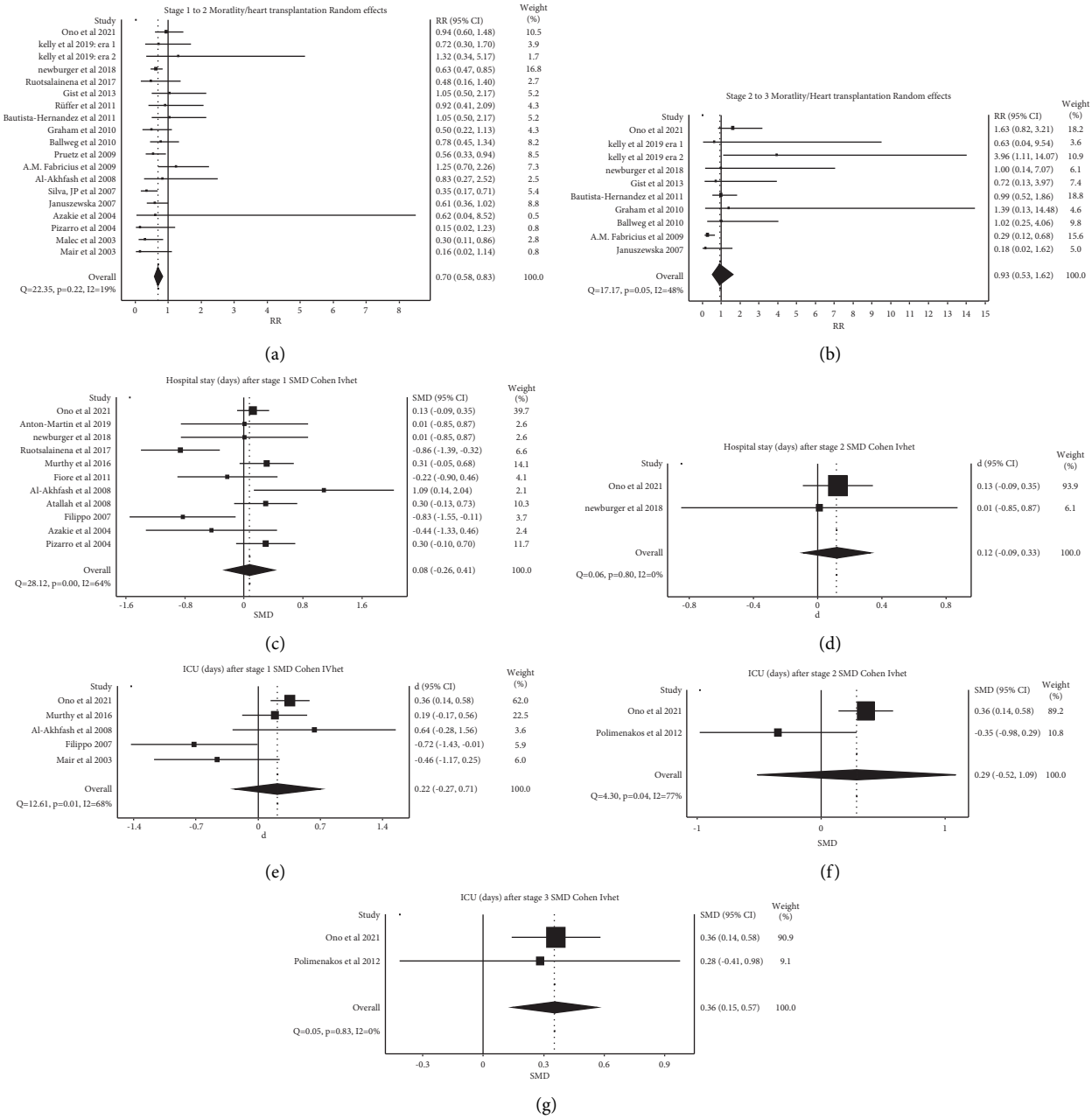

FIGURE 3: Forest plot of mortality outcomes between stage 1 and stage 2, and stage 2 and stage 3; hospital stay outcome after stage 1 and stage 2; and ICU stay after stage 1, 2, and 3. Right ventricle to pulmonary artery shunt vs. modified Blalock–Taussig shunt.

Table 5: Secondary outcomes.

Outcome	RV-PA conduit		Modified BT shunt		Study ID that reported these outcome
	Event *n* (%)	Total	Event *n* (%)	Total	
Post-Norwood complications					
Overall complications	6 (8.5%)	70	19 (17.8%)	107	34
Cardiac complications (RV dysfunction, arrhythmia, etc.)	174 (13%)	1331	157 (11.9%)	1314	5 \ 7
Respiratory complications (pleural effusion, pneumothorax, etc.)	3 (21.4%)	14	1 (5.55%)	18	68
Neurological complications (stroke, bleed, seizure, etc.)	187 (18.2%)	1027	138 (10.81%)	1276	5 \ 7
GI complications (necrotizing enterocolitis, liver failure, cirrhosis)	7 (3.9%)	177	15 (8.47%)	177	1 \ 67
ID infectious disease complications (sepsis, repertory infection, UTI, etc.)	11 (30.5%)	36	2 (8.33%)	24	49/67
Renal complications (acute or chronic renal failure)	5 (1.8%)	280	3 (1.20%)	248	22
Other complications (hematological, etc.)	5 (1.7%)	294	4 (1.50%)	266	22\ 67
Pacemaker placed	19 (2.5%)	747	23 (2.90%)	791	1\5\27
Protein-losing enteropathy	22 (3.3%)	659	10 (1.59%)	629	5\22\27
Adjudicated cause of death					
Unknown	5 (1.8%)	274	5 (1.81%)	275	5
Cardiovascular	6 (1.9%)	320	9 (2.52%)	357	5\32
Pulmonary	4 (1.5%)	274	0	275	5
Neurological	1 (0.36%)	274	0	275	5
Gastroenterologic/hepatic	1 (0.36%)	274	0	275	5
Complex/multisystem	1 (0.36%)	274	2 (0.7%)	275	5
Organ failure	N/R	N/R	N/R	N/R	N/R
Infectious	2 (0.7%)	274	0	275	5

N/R: not reported.

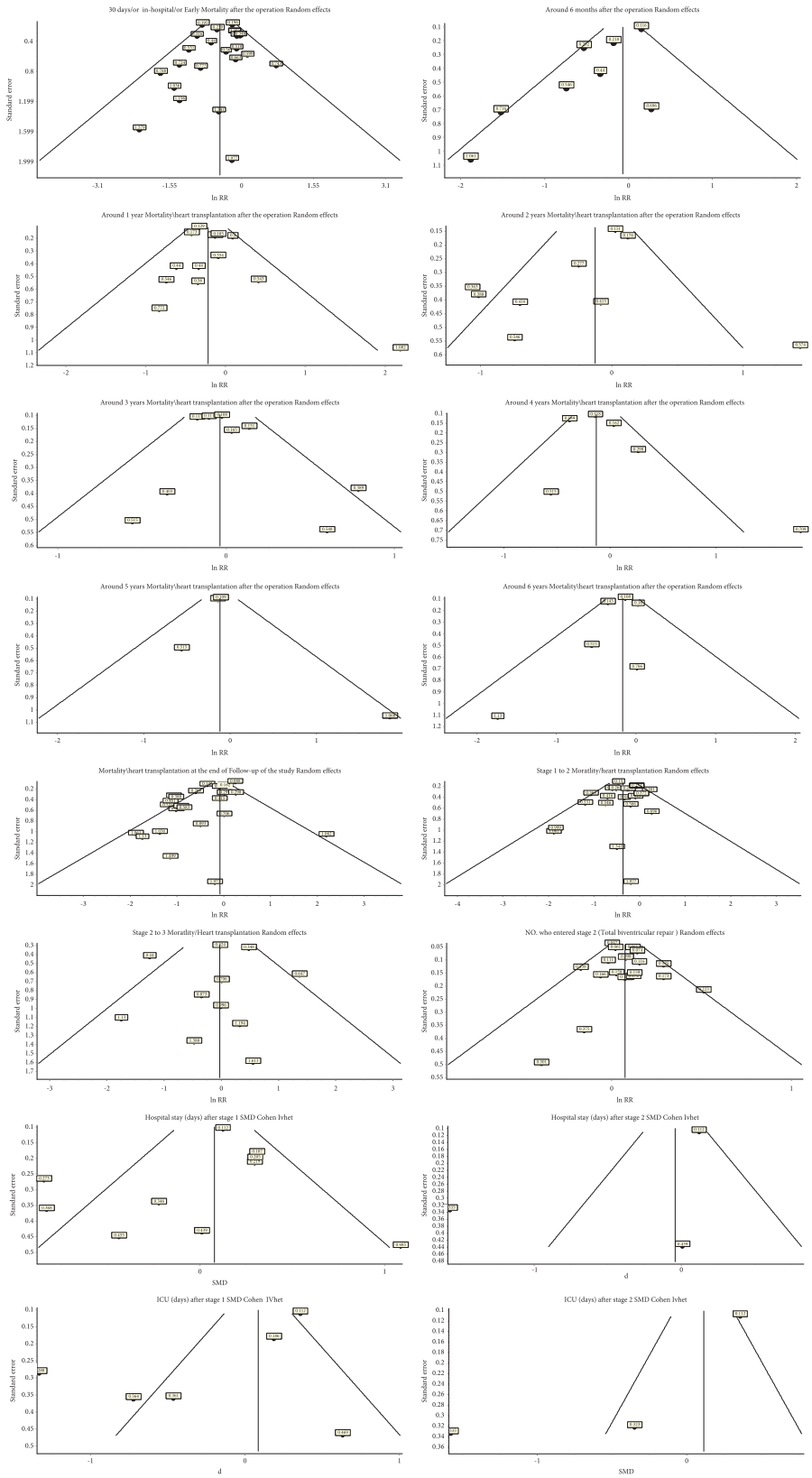

Figure 4: Funnel plots for the study's outcomes.

reoperation at S2P, and incidence of pulmonary artery stenosis prior to S2P.

5. Conclusions

The RV-PA shunt had lower mortality and overall complications rate than mBT shunt at the short-term outcome within the first two years, but at the long term, there was no difference between the two shunts. On the other hand, the mBT shunt had a lower incidence of cardiac complications at the early stage after the operations. However, some studies are poor due to the difficulties in conducting original research in this field. Therefore, we recommend conducting systematic reviews and original studies to compare these and other therapeutic procedures for these patients.

Appendix

A. Search Strategies

PICO

P: children with HLHS or single ventricle.
I: Sano shunt or its synonymous.
C: modified blalock taussig.
Primary outcome: heart transplant or death.

PubMed

Terms: ("Sano shunt" OR "initial shunt" OR "RV-to-pulmonary artery shunt" OR "Systemic-to-pulmonary artery shunt" OR "modified blalock taussig" OR "aortopulmonary shunt" OR "Norwood" OR "right ventricle-to-pulmonary artery conduit" OR "Blalock Taussig" OR "Subclavian Pulmonary Artery Shunt") AND ("hypoplastic left heart syndrome" OR "single heart" OR "univentricular heart" OR "Dominant right ventricular" OR "Left Heart Hypoplasia Syndrome")
No of results: 1306
Web of Science
Terms
Results: 1210

Embase

Terms: TITLE-ABS-KEY "Sano shunt" OR "initial shunt" OR "RV-to-pulmonary artery shunt" OR "Systemic-to-pulmonary artery shunt" OR "modified blalock taussig" OR "aortopulmonary shunt" OR "Norwood" OR "right ventricle-to-pulmonary artery Shunt" AND TITLE-ABS-KEY "hypoplastic left heart syndrome" OR "single heart" OR "univentricular heart" OR "Dominant right ventricular" OR "Left Heart Hypoplasia Syndrome".
Results: 1458

Cochrane Clinical Trials Library

Term: "Sano shunt" OR "initial shunt" OR "RV-to-pulmonary artery shunt" OR "Systemic-to-pulmonary artery shunt" OR "modified blalock taussig" OR "aortopulmonary shunt" OR "Norwood" OR "right ventricle-to-pulmonary artery conduit" OR "Blalock Taussig" OR "Subclavian Pulmonary Artery Shunt" AND "hypoplastic left heart syndrome" OR "single heart" OR "univentricular heart" OR "Dominant right ventricular" OR "Left Heart Hypoplasia Syndrome".
Results: 65

Scopus

Term: ("Sano shunt" OR "initial shunt" OR "RV-to-pulmonary artery shunt" OR "Systemic-to-pulmonary artery shunt" OR "modified blalock taussig" OR "aortopulmonary shunt" OR "Norwood" OR "right ventricle-to-pulmonary artery conduit" OR "Blalock Taussig" OR "Subclavian Pulmonary Artery Shunt") AND ("hypoplastic left heart syndrome" OR "single heart" OR "univentricular heart" OR "Dominant right ventricular" OR "Left Heart Hypoplasia Syndrome").
Results: 1948
All results: 5987

B. Funnel Plots for Each Outcome

Funnel plots for each outcome are present in Figure 4.

Data Availability

The total detailed data for the characteristics of accepted studies and patients are available from the corresponding author. For those who want to view them, they must contact the corresponding author.

Additional Points

This umbrella review was performed according to the protocol previously published on PROSPERO (CRD42021282120).

Conflicts of Interest

The authors declare that they have no conflicts of interest to report regarding the present study.

Authors' Contributions

Mr Ahmad Yamen Arnaout is the study leader and performed the screening, data extraction, analysis, quality assessment, and writing. Mr Yaman Nerabani performed the screening, data extraction, quality assessment, and writing. Mr Hassan Alhaj Ali performed the screening, data extraction, quality assessment, and writing. Mr Mohamad Zaher Shahrour performed the screening, data extraction, quality assessment, and writing. Dr Mohamad Yahia Fallaha performed the screening, data extraction, quality assessment, and writing. Mr Ibrahim Arnaout performed data extraction and writing. Mr Ahmad Sajee performed data extraction and writing. Professor Mohamad Morjan performed scientific supervising and reviewed the manuscript.

Professor Hussein Al-Kanj performed scientific supervising and reviewed the manuscript.

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