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Assessment the Accuracy of Risk Adjustment in Congenital Heart Surgery Score in Congenital Cardiac Centers at Dakahlya Government

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Article History	Abstract
Article History Received: 06 June 2023 Revised: 05 September 2023 Accepted: 21 September 2023	Abstract Background: There has been massive progress in congenital cardiac surgery outcomes analysis in the last two decades. The RACHS-1 score was developed to adjust for the huge difference in congenital cardiac pathologies when comparing the operative mortality for children undergoing paediatric congenital heart surgery. Methods: This prospective observational study was conducted on all patients undergoing cardiac surgery in all congenital heart surgery centers in Dakahlya Governorate, where we assessed the operative mortality post-surgery and compared it to the RACHS-1 expected mortality to assess the accuracy of using the RACHS-1 to predict mortality in our society. Results: This study included 559 patients in two years period, VSD was the most frequent diagnosis (13.6%), followed by Single ventricle (11.3%), then TOF (10.4%). VSD closure was the most frequent procedure (11.3%), followed by mB-T shunt (8.9%), then TOF repair (8.1%). Day-30 outcome among the studied cases Mortality was uncommon (7.9%), the main cause was heart in aetiology (43.2%). Mortality was significantly highest in HLHS, and then followed by TAPVD. The overall mortality according to each category was 1.2%, 3%, 8.57%, 17.39%, 0% and 57.1% for category 1, 2, 3, 4, 5 and 6. Conclusion: RACHS-1 is an accurate method to predict mortality early post operatively. Ventricular septal defect (VSD) was the most frequent diagnosis.
	VSD closure was the most frequent procedure, followed by mB-T shunt, then TOF repair, most of them were elective. The most common category of RACHS-
	<i>I was 3. HLHS, RACHS</i> \geq <i>3, Palliation operation were significant independent</i>
CC License	factors increase the day-30 mortality.
CC-BY-NC-SA 4.0	Keywords: Congenital, Heart, Surgery

1. Introduction

Tremendous Efforts are ongoing to improve the techniques and technologies available to evaluate and improve the outcomes of congenital cardiac surgeries. Patients after congenital cardiac surgeries are at risk of early mortality because of the abnormal cardiac function and arrhythmias, pulmonary vascular problems, and impaired lung function. They also frequently coexisting extracardiac abnormalities (Tabib et al., 2016).

The importance of outcomes analysis based on the fact that in the field of congenital cardiac surgery, using just mortality measurements without RA center to center (Cavalcante et al., 2016). There are major multi-institutional studies attempted to measure congenital cardiac surgery case complexity which are RACHS-1 score and the Aristotle Complexity Score (Jenkins et al., 2022).

RACHS-1 categories cardiac surgical procedures into six risk categories, based on expected early postoperative mortality, with category 1 representing the lowest risk and category 6 representing the highest risk. Along with the risk group, the RACHS-1 score adds age at time of surgery, prematurity, presence of a major noncardiac anomaly, and whether the patient is redo or not. The RACHS-1 score

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showed accuracy in predicting postoperative mortality in several studies in both Europe and North America (Thiagarajam & Laussen, 2015).

2. Materials and Methods

This prospective observational study was conducted on all patients undergoing cardiac surgery in all congenital heart surgery centers in Dakahlya Governorate, where we assessed the operative mortality post-surgery and compared it to the RACHS-1 expected mortality to assess the accuracy of using the RACHS-1 to predict mortality in our society over 2 years period. 559 patients were included in this study.

RACHS Category	Procedure	RACHS Category	Procedure
RACHS category 1	PDA >30 d Coarctation >30 d ASD	RACHS category 4	Truncus arteriosus repair Double switch Hypoplastic arch repair
RACHS category 2	VSD TOF Vascular ring Coarctation <30 d AP Window		Unifocalization Rastelli ASO/VSD Konno Atrial septectomy
RACHS category 3	ASO TOF/PA Ross	RACHS category 5	TV repositioning in Ebsteins anomaly <30 d Truncus/IAA
	ALCAPA DORV repair Coarctation/VSD BTS CAVC PAB	RACHS category 6	Norwood procedure DKS

Risk adjustment for congenital heart surgery (RACHS-1)

PDA indicates patent ductus arteriosus; ASD, atrial septal defect; VSD, ventricular septal defect; TOF, tetralogy of Fallot; AP, aortopulmonary; ASO, arterial switch operation; PA, pulmonary atresia; ALCAPA, anomalous left coronary artery to the pulmonary artery; DORV, double outlet right ventricle; BTS, Blalock-Tuassig shunt; CAVC, complete atrioventricular canal; PAB, pulmonary artery band; TV, tricuspid valve; DKS, Damus Kaye Stancil.

Statistical methods: The collected data were coded, tabulated, and statistically analyzed using IBM SPSS statistics (Statistical Package for Social Sciences) software version 28.0, International Business Machines Corporation., Chicago, USA, 2021. Quantitative data tested for normality using Shapiro-Wilk test, then if normally distributed described as mean±SD (standard deviation) as well as minimum and maximum of the range, then compared using independent t-test, and if not normally distributed described as Median (1st–3rd Interquartiles) then compared using Mann Whitney test. Qualitative data described as number and percentage and compared using Chi square test as well as Fisher's exact test for variables with small expected numbers. Logistic regression was used to find out factors affecting outcomes. The level of significance was taken at p-value <0.050 was significant, otherwise was non-significant.

3. Results and Discussion

 Table (1): Demographic characteristics among the studied cases

Variables		Median (1st–3rd IQ)	Range	
Age (days)	286.0 (92.0–961.0)	5.0-41399.0	
Weigh	nt (kg)	7.1 (4.1–12.0)	1.9-83.0	
		Mean±SD	Range	
GA (w	veeks)	37.5±1.4	30.0-40.0	
Birth weight (kg)		3.0±0.6	1.0-5.2	
		n	%	
Condon	Male	277	49.6%	
Gender	Female	282	50.4%	
	Children>1y	68	12.2%	
Age category	Infant<1y	491	87.8%	
Prematurity		23	4.1%	
Syndrome		50	8.9%	
Down		29	5.2%	
Heterotaxy		11	2.0%	

William's	3	0.5%
Shone's complex	2	0.4%
DiGeorge	2	0.4%
Noonan's	1	0.2%
Cutis Laxa	1	0.2%
Bardet Biedl	1	0.2%

Total=559. IQ: Interquartiles

Table (1) shows demographic characteristics among the studied cases.

Table (2): Diagnosis among the studied cases			
Diagnosis	n	%	
ALCAPA	2	0.4	
Aortic valve disease	15	2.7	
AP window	2	0.4	
ASD	35	6.3	
AVSD	23	4.1	
ccTGA	12	2.1	
СоА	30	5.4	
Coronary fistula	2	0.4	
DORV	23	4.1	
Ebsteins's anomaly	4	0.7	
HLHS	8	1.4	
Hypoplastic Aortic Arch	21	3.8	
Interrepted aortic arch	6	1.1	
LV Pseudo aneurysm	1	0.2	
Mitral valve disease	21	3.8	
PA, IVS	5	0.9	
PA, VSD	11	2.0	
PA, VSD, MAPCAs	5	0.9	
PAPVR	3	0.5	
PAVSD	15	2.7	
PDA	16	2.9	
RVOTO	11	2.0	
SAM	4	0.7	
Single ventricle	63	11.3	
TAPVD	10	1.8	
TGA, IVS	37	6.6	
TGA, VSD	10	1.8	
TGA, VSD, PS	10	1.8	
TOF	58	10.4	
Truncus Arteriosus	13	2.3	
Vasclar ring	7	1.3	
VSD	76	13.6	

Table (2) shows VSD was the most frequent diagnosis (13.6%), followed by Single ventricle (11.3%), then TOF (10.4%)

Table (3): Procedure among the studied cases		
2	0.4	
9	1.6	
2	0.4	
27	4.8	
36	6.4	
33	5.9	
	2 9 2 27 36 33	

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AVSD Repair	14	2.5	
BDG	24	4.3	
Bilateral PA Banding	1	0.2	
CoA repair	30	5.4	
Cone repair	4	0.7	
CORONARY artery fistula closure	2	0.4	
DORV Repair	16	2.9	
Double switch	8	1.4	
Fontan	12	2.1	
LV aneurysm repair	1	0.2	
M Aortic valve replacement	1	0.2	
M Mitral replacement	6	1.1	
mBT shunt	50	8.9	
Mitral Valve repair	15	2.7	
modified-Konno	1	0.2	
Mustard	7	1.3	
Nikaidoh	5	0.9	
Norwood	7	1.3	
PA banding	39	7.0	
PA, VSD repair	8	1.4	
PAPVR repair	3	0.5	
PAVSD repair	14	2.5	
PDA Ligation	16	2.9	
Rastelli	3	0.5	
Ross	5	0.9	
RV-PA conduit placement	1	0.2	
RVOTO repair	9	1.6	
SAM resection	3	0.5	
Senning	3	0.5	
TAPVD repair	10	1.8	
TOF repair	46	8.3	
Truncus arteriosus repair	12	2.1	
Unifocalization	3	0.5	
Vascular ring division	7	1.3	
VSD closure	63	11.3	
Yasui Operation	1	0.2	

Table (3) shows VSD closure was the most frequent procedure (11.3%), followed by mB-T shunt (8.9%), then TOF repair (8.1%).

Table (4): Day-30 outcome among the studied cases			
Ţ	Variables		%
Montolity	Death	44	7.9%
Mortanty	Survival	515	92.1%
	Neurological complications	4	9.1%
	Cardiac complications	19	43.2%
Causes of mortality	Pulmonary complications	10	22.7%
	MOF	7	15.9%
	Shunt Problems	4	9.1%

Table (4) shows **Day-30 outcome among the studied cases**. Mortality was uncommon (7.9%), the main cause was heart in aetiology (43.2%).

Varia	ables	Mortality (n=44)	Survival (n=515)	p-value
Age (Davs)		59.0	299.0	∩0.002 *
		(25.0–954.0)	(110.0–973.0)	20000
Weigh	ıt (kg)	4.1 (3.0–11.1)	7.5 (4.5–12.0)	△0.002 *
Condon	Male	17 (38.6%)	260 (50.5%)	#0 121
Gender	Female	27 (61.4%)	255 (49.5%)	#0.151
GA (w	veeks)	37.1±1.2	37.5±1.4	^0.057
Birth wei	ight (kg)	2.8±0.5	3.0±0.6	^0.056
Prema	turity	3 (6.8%)	20 (3.9%)	§0.414
Syndi	rome	3 (6.8%)	47 (9.1%)	§0.787
Dov	wn	1 (2.3%)	28 (5.4%)	§0.719
Heter	otaxy	1 (2.3%)	10 (1.9%)	§0.598
Willia	am's	0 (0.0%)	3 (0.6%)	§0.999
Shone's o	complex	0 (0.0%)	2 (0.4%)	§0.999
DiGe	orge	1 (2.3%)	1 (0.2%)	§0.151
Noon	an's	0 (0.0%)	1 (0.2%)	§0.999
Cutis	Laxa	0 (0.0%)	1 (0.2%)	§0.999
Bardet	Biedl	0 (0.0%)	1 (0.2%)	§0.999

 Table (5): Comparison showing effect of demographic characteristics on day-30 mortality

^Independent t-test. △Mann Whitney test. #Chi square test. §Fisher's Exact test. *Significant Table (5) shows Cases dead by day-30 significantly had lower age and weight.

	Mortality	Survival
Diagnosis	(n=44)	(n=515)
ALCAPA	1 (50.0%)	1 (50.0%)
Aortic stenosis	0 (0.0%)	1 (100.0%)
Aortic valve disease	0 (0.0%)	14 (100.0%)
AP window	0 (0.0%)	2 (100.0%)
ASD	1 (2.9%)	34 (97.1%)
AVSD	1 (4.3%)	22 (95.7%)
ccTGA	2 (16.7%)	10 (83.3%)
СоА	0 (0.0%)	30 (100.0%)
Coronary fistula	0 (0.0%)	2 (100.0%)
DORV	1 (4.3%)	22 (95.7%)
DTGA	0 (0.0%)	1 (100.0%)
Ebsteins's anomaly	0 (0.0%)	4 (100.0%)
HLHS	4 (57.1%)	3 (42.9%)
Hypoplastic Aortic Arch	2 (9.5%)	19 (90.5%)
Interrepted aortic arch	0 (0.0%)	6 (100.0%)
LV aneurysm	0 (0.0%)	1 (100.0%)
Mitral valve disease	2 (9.5%)	19 (90.5%)
PA, IVS	1 (20.0%)	4 (80.0%)
PA, VSD	2 (18.2%)	9 (81.8%)
PA, VSD, MAPCAs	0 (0.0%)	5 (100.0%)
PAPVR	0 (0.0%)	3 (100.0%)
PAVSD	1 (6.7%)	14 (93.3%)
PDA	0 (0.0%)	16 (100.0%)
RVOTO	0 (0.0%)	11 (100.0%)
SAM	0 (0.0%)	4 (100.0%)
Single ventricle	9 (14.8%)	54 (85.2%)
TAPVD	3 (33.3%)	7 (66.7%)

 Table (6): Comparison according to day-30 mortality regarding Diagnosis

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TGA, IVS	4 (11.1%)	32 (88.9%)
TGA, VSD	1 (10.0%)	9 (90.0%)
TGA, VSD, PS	0 (0.0%)	10 (100.0%)
TOF	3 (5.2%)	55 (94.8%)
Truncus Arteriosus	3 (23.1%)	10 (76.9%)
Vasclar ring	0 (0.0%)	7 (100.0%)
VSD	2(2.6%)	74 (97,4%)

VSD 2 (2.6%) 74 (97.4%) Percentages were taken from rows (per diagnosis). §Fisher's Exact test. *Significant Table (6) shows Mortality was significantly highest in HLHS, then followed by TAPVD

Table (7): Comparison according to a	Table (7): Comparison according to day-30 mortality regarding procedure				
Procedure	Mortality	Survival	n-value		
	(n=44)	(n=515)	p value		
ALCAPA repair	1 (50.0%)	1 (50.0%)			
Aortic valve repair	0 (0.0%)	9 (100.0%)			
AP window repair	0 (0.0%)	2 (100.0%)			
Arch reconstruction	2 (7.4%)	25 (92.6%)			
Arterial switch	3 (8.3%)	33 (91.7%)			
ASD closure	1 (3.0%)	32 (97.0%)			
AVSD Repair	1 (7.1%)	13 (92.9%)			
BDG	1 (4.2%)	23 (95.8%)			
Bilateral PA Banding	0 (0.0%)	1 (100.0%)			
CoA repair	0 (0.0%)	30 (100.0%)			
Cone repair	0 (0.0%)	4 (100.0%)			
CORONARY artery fistula closure	0 (0.0%)	2 (100.0%)			
DORV Repair	1 (6.3%)	15 (93.8%)			
Double switch	1 (12.5%)	7 (87.5%)			
Fontan	4 (33.3%)	8 (66.7%)			
LV aneurysm repair	0 (0.0%)	1 (100.0%)			
M Aortic valve replacement	0 (0.0%)	1 (100.0%)			
M Mitral replacement	1 (16.7%)	5 (83.3%)			
mBT shunt	7 (14.0%)	43 (86.0%)			
Mitral Valve repair	1 (6.7%)	14 (93.3%)	8~0 001*		
modified-Konno	0 (0.0%)	1 (100.0%)	8<0.001		
Mustard	0 (0.0%)	7 (100.0%)			
Nikaidoh	0 (0.0%)	5 (100.0%)			
Norwood	4 (57.1%)	3(42.9%)			
PA banding	2 (5.1%)	37 (94.9%)			
PA, VSD repair	1 (12.5%)	7 (87.5%)			
PAPVD repair	0 (0.0%)	3 (100.0%)			
PAVSD repair	1 (7.1%)	13 (92.9%)			
PDA Ligation	0 (0.0%)	16 (100.0%)			
Rastelli	0 (0.0%)	3 (100.0%)			
Ross	0 (0.0%)	5 (100.0%)			
RV-PA conduit placement	0 (0.0%)	1 (100.0%)			
RVOTO repair	0 (0.0%)	9 (100.0%)			
SAM resection	0 (0.0%)	3 (100.0%)			
Senning	1 (33.3%)	2 (66.7%)			
TAPVD repair	3 (30.0%)	7 (70.0%)			
TOF repair	2 (4.4%)	44 (95.6%)			
Truncus arteriosus repair	3 (25.0%)	9 (75.0%)			
Unifocalization	0(0.0%)	3 (100.0%)			
Vascular ring division	0(0.0%)	7 (100.0%)			

 Table (7): Comparison according to day-30 mortality regarding procedure

VSD closure	2 (3.2%)	61 (96.8%)
Yasui Operation	0 (0.0%)	1 (100.0%)

Percentages were taken from rows (per procedure). §Fisher's Exact test. *Significant Table (7) shows Mortality was significantly highest in Norwood, followed by Fontan 33.3% then TAPVD repair 30%.

Variables	•	Mortality Survival		 	
variables		(n=44)	(n=515)	p-value	
Urgency	Elective	29 (65.9%)	406 (78.8%)		
	Emergency	3 (6.8%)	18 (3.5%)	#0.129	
	Urgent	12 (27.3%)	91 (17.7%)		
Procedure type	Repair	25 (56.8%)	383 (74.4%)		
	Palliation	19 (43.2%)	132 (24.7%)	§0.030 *	
RACHS	1	1 (2.3%)	82 (15.9%)		
	2	5 (11.4%)	158 (30.7%)		
	3	18 (40.9%)	192 (37.3%)	§<0.001 *	
	4	16 (36.4%)	76 (14.8%)		
	5	0 (0.0%)	4 (0.8%)		
	6	4 (9%)	3 (0.6%)		

 Table (8): Comparison according to day-30 mortality regarding Operation characteristics

#Chi square test. §Fisher's Exact test. *Significant

Table (8) shows Cases dead by day-30 significantly had more frequent palliation and RACHS 3. Calculating overall mortality according to each category was 1.2%, 3%, 8.57%, 17.39%, 0% and 57.1% for category 1, 2, 3, 4, 5 and 6

		8 20	
Variab	les	n	%
	Elective	435	77.8%
Urgency	Emergency	21	3.8%
	Urgent	103	18.4%
Procedure type	Repair	408	73.0%
	Palliation	151	27%
RACHS score	1	83	14.8%
	2	163	29.2%
	3	210	37.6%
	4	92	16.5%
	5	4	0.7%
	6	7	1.3%

 Table (9): Operation characteristics among the studied cases

Table (9) shows **Operation characteristics among the studied cases**. Operations were mainly elective (77.8%) and patients for repair (73.0%).

Table (10): Regression model for factors affecting day-30 mortality

Factors	β	SE	p-value	OR (95% CI)
HLHS	1.84	0.83	0.027*	6.30 (1.23–32.12)
RACHS≥3	1.01	0.49	0.039*	2.73 (1.05-7.12)
Palliation operation	0.84	0.38	0.027*	2.32 (1.10-4.91)
Constant	-3.92	0.43	<0.001*	

 β : Regression coefficient. SE: Standard error. OR: Odds ratio. CI: Confidence interval. *Significant HLHS, RACHS \geq 3, Palliation operation were significant independent factors increase the likelihood of 30-day mortality.

This prospective observational study was conducted on 559 patients underwent cardiac surgery in all congenital heart surgery centers in Dakahlya Governorate, which were followed for 30 days as regards operative mortality.

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Comparing our results to the RACHS-1 we showed mortality of 1.2%, 3%, 8.57%, 17.39%, 0% and 57.1% for category 1, 2, 3, 4, 5 and 6, respectively while The RACHS-1 score ranges from 1–6 (mortality rates were 0.4% in category 1, 3.8% in 2, 8.5% in 3, 19.4% in 4, and 47.7% in 6) so we showed higher mortality in both category 1 and 6 otherwise the rest of the results were very similar which mean we had a very good outcome that could be compared to the standard care worldwide even better than other big centers as shown in a big Brazilian study which showed a mortality of 1.8%, 5.5%, 14.9%, 32.5% and 68.6% for category 1, 2, 3, 4 and 6, respectively (Cavalcante et al., 2016).

Regarding demographic data, the median (1st-3rd IQ) age of the studied group was 286.0 (92.0–961.0) Days, most of them were infants (87.2%) and females (50.4%). The median (1st-3rd IQ) weight was 7.1 (4.1–12.0) Kg. Only 5.2% of patients were Down syndrome.

While; a Brazilian retrospective date analysis done by **Cavalcante et al., (2016)** that were conducted on 3201 surgeries. Of these, 3071 were classified according to the RACHS-1 score and 130 procedures could not be categorized by the score. Of the patients, 1643 (51.7%) were male, aged less than 1 year of age 1523 (47.5%) and 2281 (71.1%) used CPB. The median weight was 8 kg (4 to 17.8 kg). Only 5.5% had trisomy 21.

As regard diagnosis among the studied cases, the current study detected that ventricular septal defect (VSD) was the most frequent diagnosis (13.6%), followed by Single ventricle (11.3%), then tetralogy of Fallot (TOF) (10.4%).

An investigation used the cardiovascular surgery patients in the Trichotomous Outcome Prediction in Critical Care database collected by the Child Health and Human Development Collaborative Pediatric Critical Care Research Network. The overall sample contained 10,078 patients, of whom 1550 underwent a cardiac surgery. Of the cardiac interventions, 1199 (77.4%) had 2-ventricle anatomy and 351 (22.6%) were single-ventricle patients. A total of 871 (56.2%) were acyanotic and 679 (43.8%) were cyanotic (Berger et al., 2017).

Concerning procedure among the studied cases, VSD closure was the most frequent procedure (11.3%), followed by mB-T shunt (8.9%), then TOF repair (8.1%). Operations were mainly elective (77.8%), for total repair (73.0%). Furthermore, the most common category of RACHS was 3 (21037.6%).

With reference to Day-30 outcome among the studied cases, mortality was uncommon (7.9%), the main cause was heart failure in etiology (43.2%).

According to NATIONAL CONGENITAL HEART DISEASE AUDIT (NCHDA), in Newcastle Freeman Hospital, 30-day mortality for RACHS-1 1 and 2 categories were 2.4% in patient > 16 yrs underwent congenital heart procedure. While in Leicester Glenfield Hospital, it was 1.9%. The 30-day mortality rate in Glasgow Royal Hospital for Children was 1.5%. Bristol Royal Hospital for Children, Southampton Wessex Cardiothoracic Centre, Dublin Our Lady's Children's Hospital and Liverpool Alder Hey Hospital revealed similar rate of mortality. Likely, **Miyata et al. (2014)** reported that outcome rates of congenital heart surgery were 1.7% for 30-day mortality.

A satisfactory 30-day mortality rate can be attributed to the adequate provision of resources for hospitalization care, such as postoperative intensive care, and is reflected by long-term hospitalization that lasts on average one month because mortality might be related with prolonged stay.

Comparison according to day-30 mortality, cases dead by day-30 significantly had lower age and weight. Mortality was significantly highest in HLHS, then ALCAPA also this result is not accurate as we had only 2 cases, followed by TAPVD, Mortality was significantly highest in Norwood (57.1%), followed by Fontan and Senning (33.3% for each). Cases dead by day-30 significantly had more palliative procedures and RACHS 3. So, HLHS, RACHS \geq 3, Palliation operation and Open surgery were significant independent factors increase the likelihood of day-30 mortality.

The National Institute for Cardiovascular Outcomes Research (NICOR) and audit at UK regarding Norwood procedure in HLHS quotes a 33% mortality early post-operatively and that was in Birmingham Children's Hospital which is one of the biggest centers who dealt with HLHS with great success up to Fontan and 45% mortality between stage one and two so our result of 57% mortality

early postoperatively is not a bad result compared to the biggest centers who deal with HLHS and this is for many reasons, **firstly** this is the start of a learning curve which shows good result compared to the 100% mortality of HLHS if left not treated, **secondly** in centers who don't do this procedure regularly our results almost the same in spite of the availability of ECMO and NO in these centers, **thirdly** in Birmingham children hospital we used to classify HLHS into 2 groups with one group who has high risk of mortality due to multiple risk factors which include ; severe TR, restrictive atrial septum and very small ascending aorta less than 2mm, In our study 2 of the cases who died were from the high risk group so I believe with better selection of the cases and more cases to be done this will improve the learning curves and eventually the results.

Study limitation

There were some limitations that had to be disclosed in this study. **First**, this study was done in centers in Dakahlya only and we need to involve more centers all across Egypt to get more accurate results that we can use to formulate a model that can be generalized all across Egypt. This would require a comprehensive database across multiple institutions to accurately capture all essential variables. **Second**, we did not compare RACHS 1 with other risk stratification score in cardiac surgery to detect its efficiency. **Finally**, the sample size of this study was relatively small in comparison to similar study. The number of cases in category 5 AND 6 are small so in order to have more accurate results we need more cases although these 2 categories have rare cases also. One of the limitations also was that 20 patients were lost in follow up.

4. Conclusion

RACHS-1 is an accurate method to predict mortality early post operatively. Ventricular septal defect (VSD) was the most frequent diagnosis. VSD closure was the most frequent procedure, followed by mB-T shunt, then TOF repair, most of them were elective. The most common category of RACHS was 3. 30-day mortality was 7.9%. Patients dead by day-30 had lower age and weight. Mortality was significantly highest in HLHS, followed by TAPVD; Mortality was significantly highest in Norwood, followed by Fontan and Senning. Cases dead by day-30 significantly had more frequent palliation and RACHS \geq 3. HLHS, RACHS \geq 3, Palliation operation were significant independent factors increase the day-30 mortality.

References:

- Tabib, A., Abrishami, S.E., Mahdavi, M. (2016). Predictors of Prolonged Mechanical Ventilation in Pediatric Patients after Cardiac Surgery for Congenital Heart Disease. *Research in Cardiovascular Medicine*, 5(3). <u>https://doi.org/10.5812/cardiovascmed.30391</u>.
- Cavalcante, C.T. de M.B., de Souza, N.M.G., Pinto Júnior, V.C. (2016). Analysis of surgical mortality for congenital heart defects using RACHS-1 risk score in a Brazilian single center. *Brazilian Journal of Cardiovascular Surgery*, 31(3), 219–225. https://doi.org/10.5935/1678-9741.20160022.
- Jenkins, K.J., Koch Kupiec, J., Owens, P.L. (2022). Development and Validation of an Agency for Healthcare Research and Quality Indicator for Mortality After Congenital Heart Surgery Harmonized With Risk Adjustment for Congenital Heart Surgery (RACHS-1) Methodology. *Journal of the American Heart Association*, 5(5), e003028. <u>https://doi.org/10.1161/JAHA.115.003028</u>.
- Thiagarajan, R.R., & Laussen, P.C. (2015). Risk Adjustment for Congenital Heart Surgery -1 (RACHS-1) for Evaluation of Mortality in Children Undergoing Cardiac Surgery. In Barach, P.R., Jacobs, J.P., Lipshultz, S.E., et al. (Eds.), *Pediatric and Congenital Cardiac Care: Volume 1: Outcomes Analysis* (pp. 327–336). Springer London. <u>https://doi.org/10.1007/978-1-4471-6587-3_26</u>.
- Berger JT, Holubkov R, Reeder R, Wessel DL, Meert K, Berg RA, et al. (2017). Physiological Profiles and Surgical Complexity. *The Journal of Thoracic and Cardiovascular Surgery*, 154(2), 620-8.
- National Congenital Heart Disease Audit (NCHDA). (2021). *NCHDA Domain Report 2021*. Retrieved from <u>https://www.nicor.org.uk/wp-content/uploads/2021/10/NCHDA-Domain-Report_2021_FINAL.pdf</u>.
- Miyata H, Murakami A, Tomotaki A, Takaoka T, Konuma T, Matsumura G, et al. (2014). Predictors of 90-day mortality after congenital heart surgery: the first report of risk models from a Japanese database. *The Journal of Thoracic and Cardiovascular Surgery*, 148(5), 2201-6.
- Welsby IJ, Bennett-Guerrero E, Atwell D, White WD, Newman MF, Smith PK, et al. (2002). The association of complication type with mortality and prolonged stay after cardiac surgery with cardiopulmonary bypass. *Anesthesia & Analgesia*, 94(5), 1072-8.