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Original article

Congenital cardiac anomalies and imperforate anus: A hospital's experience

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ABSTRACT

Objectives: We aimed in our study to determine the incidence and type of congenital cardiac anomaly (CCA) in newborns associated with imperforate anus (IA), the outcome of surgical reconstruction of the anorectum in our center, and the need for performing echocardiography in all patients with IA.
Materials and methods: The preoperative echocardiography reports of all cases born with IA and managed at King Abdulaziz University Hospital, Jeddah, Saudi Arabia over a period of 11 years (Jan 2000–Dec 2010) were reviewed. The average annual delivery rate of this hospital is 5500.
Results: During the study period, 61 patients of IA were diagnosed that showed an incidence of about 1 per 992 live births, and the rate of CCA among the IA subjects was 15 (24.6%). In 12 patients (19.6%), the associated CCAs were of a mild nature, and reconstruction of the anorectum went smoothly. Three patients (4.9%) had significant CCA and died.
Conclusion: The incidence of IA in our hospital is 1 per 992 live births, and its association with CCA is 24.6%. The majority of CCAs associated with IA were of the mild type.

1. Introduction

Imperforate anus (IA) is a major form of anorectal malformations or 'ARM' in which the baby is born without a normal anal opening.[1] The incidence of anorectal malformations (ARMs) varies from 2 to 2.5 per 10,000 live births, with significant variations in the prevalence between regions throughout the world.[2] IA is not a lethal anomaly; however, the management of patients with these malformations can be complex, especially with the high frequency of associated anomalies that ranges from 40% to 70% in ARM patients.[3–7] Most of the time, the morbidity and mortality among ARM patients are due to associated anomalies that give an indication for compulsory comprehensive evaluation of these patients.[5,8–10] As mentioned in the study of Bhargawa, among the anomalies associated with ARM, urogenital is the commonest followed by defects of spine, extremities, and cardiovascular system.[11] Cardiac anomalies occur in 9–20% of the patients with ARMs, with Tetralogy of Fallot (TOF) being the commonest.[2]

The majority of associated cardiac lesions are corrected after repair of the anomaly except the complex variety with cyanosis.[12] Although complex congenital cardiac anomalies (CCAs) are obvious from the presence of cyanosis or signs of heart failure, the exact diagnosis of these lesions and of the minor ones can only be made by proper echocardiographic evaluation.[13]

The incidence and type of cardiac anomalies associated with IA have not been reported or available in Saudi Arabia; so, we aimed in our study to determine the incidence and type of associated CCAs in newborns with IA, the outcome of surgical reconstruction of the anorectum in our center, and the need for performing echocardiography in all patients with IA.

2. Materials and methods

The preoperative echocardiography reports of all cases with IA managed at King Abdulaziz University Hospital, Jeddah, Saudi Arabia over a period of 11 years (Jan 2000–Dec 2010) were reviewed. The average annual delivery rate of this hospital is 5500. The cases that were delivered in our hospital with IA and CCA were included as study subjects. The findings were tabulated with the gender of the patient, type of IA, other associated anomalies, and the type of surgery performed. Moreover, the symptoms and severity of cardiac involvement were also determined.

Data was analyzed by using Microsoft excel version 7 on a personal computer and subjected to descriptive analysis.

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Table 1
Asymptomatic patients with positive echo findings (survived).

S/N	Gender	Type of IA	Associated anomalies	Associated cardiac anomalies (echocardiography findings)	Procedure
1	Male	Recto-urethral	Esophageal atresia	TOF, PDA, ASD	Staged PSARP
2	Male	Recto-urethral	None	TOF	Staged PSARP
3	Male	Recto-urethral	None	PFO, PDA	Staged PSARP
4	Male	Recto-urethral	None	ASD, VSD	Staged PSARP
5	Male	Recto-vesical	None	PDA	Staged PSARP
6	Male	High, no fistula	Down's syndrome, vesico-ureteral reflux	TOF, ASD, VSD	Staged PSARP
7	Male	Recto-urethral	Hemivertebra	Dextrocardia, ASD	Staged PSARP
8	Female	Perineal	None	PDA	ASARP
9	Female	Recto-vestibular	None	MR, TR	ASARP
10	Female	Perineal	None	VSD, TS, PS	ASARP
11	Female	Recto-vestibular	None	VSD	ASARP
12	Female	Perineal	None	TR	ASARP

IA = Imperforate anus, TOF = Tetralogy of Fallot, PDA = Patent ductus arteriosus, ASD = Atrial septal defect, PFO = Patent foramen ovale, VSD = Ventricular septal defect, MR = Mitral regurgitation, TR = Tricuspid regurgitation, TS = Tricuspid stenosis, PS = Pulmonary stenosis, PSARP = Posterior sagittal anorectoplasty, ASARP = Anterior sagittal anorectoplasty.

2.1. Ethical issues

This study was approved by the research ethical committee of King Abdulaziz University Hospital, Jeddah.

3. Results

A total of 61 patients with various forms of IA were diagnosed and managed (32 males and 29 females); all had echocardiography as routine preoperative evaluation. The incidence of IA was about 1 per 992 deliveries, and the rate of CCA among IA subjects was 15 (24.6%), that is, nine males and six females. In 12 patients (19.6%), the associated CCAs were of a mild nature and reconstruction of the anorectum went smoothly. Three patients (4.9%), had significant CCAs and died (two postoperative and one with Edwards syndrome who was not operated) [Tables 1 and 2].

4. Discussion

The exact incidence of IA in Saudi Arabia is not known. In Asser region (southern part of Saudi Arabia), IA constituted 44.8% of all gastrointestinal malformations seen over a period of 6 years.[14] The incidence of IA has been reported in our hospital as 1 per 992 deliveries that is higher than that reported in Canada, that is, 1 per 2425 live births,[4] which might be due to mixed nationalities living in Jeddah, and the high rate of consanguineous marriages; our hospital provides services to all people, irrespective of their nationalities.

IA and its association with one or more anomalies can be corrected in a single or in multiple stages starting from the neonatal period.[15] The mild forms like perineal fistula and the covered anus type are usually corrected immediately, whereas the rest are done in stages.[16,17] Neonates with IA involve surgical procedures that are particularly affected by gestational age, birth weight, duration of the procedure, and other associated anomalies particularly cardiac anomalies. Based on the severity and hemodynamic effect, medical or surgical management of the CCA may be needed before the surgical repair of IA.[18]

In the two patients who died, surgical correction of the CCA was not possible in our hospital as pediatric cardiac surgery service was not established at that time; however, both patients were on anti-failure medications but they could not tolerate the postoperative hemodynamic changes.

The association between CCA and gastrointestinal malformations is known; Chehab et al from Lebanon reported the occurrence of CCA in 38% of 105 patients with gastrointestinal malformations. Olgun et al from Turkey reported 23.7% incidence of CCA among cases of IA,[19,20] whereas we also have the same, that is, 24.6%. A study in Iran revealed 50.4% incidence rate of CCA in newborns with IA.[1]

Voisin et al in 1978 reported that children with congenital heart disease have a much higher incidence of intestinal malformations than those with a normal heart. Among these patients, the incidence of congenital heart diseases is 9–14%, with a predominance of ventricular septal defect (VSD) and TOF.[21]

The majority of CCAs do not cause cyanosis or cardiac failure and their correction can be delayed till after correction of the anal problem; however, some of the cardiac anomalies are complex and may need some medications like prostaglandin as in cases with ductal-dependent lesions while the anal procedure is being done. Anomalies like transposition of the great arteries may need palliative procedures such as atrial septostomy to improve atrial blood mixing; this is one of the conditions where the cardiac lesion should be dealt with before the repair of IA.[12]

In our series of 61 cases, all were of the major group and echocardiography was done as part of the routine preoperative evaluation. Positive findings was seen in 15, whereas three had significant CCA, the first with Edward's syndrome and cardiomegaly which was incompatible with life and he died on the third day of life; the other two were operated, and one had colostomy and the other had anoplasty of a mild perineal form. They died because of their unstable hemodynamic status [the first had large atrial septal defect (ASD) and severe tricuspid regurgitation; the second had large ASD and large VSD]. Twelve patients had mild CCA with insignificant hemodynamic effect and had their anal problem corrected similar to those without CCA. One patient had successful correction of his TOF after completion of a three-staged correction of rectourethral type; the rest did not require correction of their cardiac anomaly.

Table 2
Symptomatic patients with positive echo findings (died).

S/N	Gender	Type of IA	Echo findings	Associated anomaly	Timing of mortality
1	Male	High, no fistula	Cardiomegaly, large PDA, VSD, PS	Trisomy 18 (Edwards syndrome)	Before surgery
2	Male	High, no fistula	Large ASD and VSD	Esophageal atresia and TOF, UDT, jaundice	After emergency colostomy
3	Female	Ano-perineal	Large ASD and VSD	Cleft Lip and Palate	After anoplasty

IA = Imperforate anus, PDA = Patent ductus arteriosus, ASD = Atrial septal defect, VSD = Ventricular septal defect, PS = Pulmonary stenosis, TOF = Tetralogy of Fallot, UDT = Undescended testicles.

5. Conclusion

The incidence of IA in our hospital is 1 per 992 live births, which is higher than the international figures; its association with CCA is 24.6%. The majority of CCAs associated with IA were of the mild type and had no effect on the surgical reconstruction of the anus, routine preoperative echocardiography evaluation of all cases of IA particularly males with the high types is important for better outcome.

Conflicts of interest

All authors have none to declare.

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King Abdulaziz University Hospital, Jeddah, KSA.

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