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Staged Reconstruction Surgeries And Fetal Interventions In Hypoplastic Right Heart Syndrome And Hypoplastic Left Heart Syndrome – When Are These Surgeries Done And How Many Actually Undergo These Surgeries?

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Abstract

Objectives

Hypoplastic right heart syndrome and Hypoplastic left heart syndrome are congenital heart diseases affecting the right and left side of the heart respectively. 3 procedures are performed for the staged reconstruction surgeries for these syndromes: Norwood, Glenn, Fontan. Fetal cardiac interventions in the form of fetal aortic balloon valvuloplasty and fetal pulmonary balloon valvuloplasty are available. In this study I analyze the ages at which the patients undergo such surgeries and how many patients undergo these surgeries.

Methods

A google survey form was prepared with questions on the reconstruction heart surgeries where participants were asked on the diagnosis, number of surgeries undergone, the age at which patients underwent such surgeries, and whether they were offered the option of Fetal intervention and if they did undergo Fetal Intervention.

Results

A total of 28 responses were received of which 22 were for HLHS while 6 were for HRHS. It was found that out of all the responses only 1 response stated that fetal intervention was done while 3 responses stated that they did receive the option for fetal intervention. Another observation which was noted is that all but one participant stated that one more reconstruction surgeries were done. A lot of these surgeries were performed within the time frame in which doctors recommend doing these surgeries.

Conclusions

Staged reconstruction surgeries are widely known today but it's more difficult counterpart , Fetal Intervention is much more rare which has lead to doctors not suggesting this procedure.

Keywords – Hypoplastic left heart syndrome(HLHS), Hypoplastic right heart syndrome(HRHS), Norwood, Glenn, Fontan, Fetal interventions.

I. Background

Congenital heart defects are the most common heart defects and Hypoplastic Right Heart Syndrome(HRHS) and Hypoplastic Left Heart Syndrome(HLHS) are two of the many heart defects. The cardiac surgeries for both of these conditions consists of 3 steps: The Norwood procedure, the Glenn procedure and the Fontan procedure; however a child may skip one of them. Together the three of them are called the staged reconstruction surgeries

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The Norwood procedure is the first and the most complex of the three. It is usually performed in in the first weeks of life after a baby is born.

In the Norwood procedure for Hypoplastic right heart syndrome the Blalock–Thomas–Taussig shunt (commonly called the Blalock–Taussig shunt)is used[1]. The goals of such a procedure is to increase blood flow to lungs, maintain adequate preload and maintain afterload by. Alfred Blalock and Helen Taussig described the original BT Shunt in which There is end-to-side anastomosis between the subclavian (or the innominate) and the pulmonary arteries. the operation was performed on the side opposite to the aortic arch to reduce kinking of the subclavian artery as it crosses over the aortic prominence. In addition, kinking is reduced by longer innominate artery(2). Another shunt called the modified Blalock- Taussig shunt is the most commonly performed procedure and it involves the use of interposition PTFE graft between the subclavian and the pulmonary artery[2]. Both the shunts have their own set of pros and cons.

The Norwood surgery is one of the most complex and high risk procedures in the staged reconstruction surgeries for Hypoplastic left heart syndrome[3]. The Norwood procedure has 3 requirements which haven't changed from dr. Norwood's initial explanation:

- There should be unobstructed systemic outflow from the single right ventricle to a reconstructed aorta,
- There should also be unobstructed pulmonary venous return into the right atrium,
- The last one is controlled pulmonary blood flow (PBF)[4].

Today there are 2 sources for the pulmonary blood flow: In the classic Norwood procedure, a modified Blalock-Taussig shunt (MBTS) provides PBF from the innominate or subclavian artery to the pulmonary arteries via a polytetrafluoroethylene tube(4), With a Sano shunt, an incision is made in the wall of the single ventricle, and a Gore-Tex conduit is used to connect the ventricle to the pulmonary artery. Direct canalization to the right ventricle provides pulsatile blood flow compared to the Blalock-Taussig conduit[5].

The Glenn procedure forms the 2nd stage of the reconstruction surgeries. A classic Glenn's shunt is a connection between the superior vena cava and right pulmonary artery. Fine sutures are used to connect the superior vena cava to the right branch of pulmonary artery and the pulmonary artery is divided or tied up, this is what is called the bidirectional shunt. In very small babies especially those aged less than 2 years , in whom lung vessel resistance is still quite high, the bidirectional Glenn's shunt is preferred. it is also preferred in borderline cases with abnormal pulmonary arteries. While avoiding the risk of failure of a complete Fontan operation, it also partly relieves symptoms[6].

The last surgery used in the staged reconstruction surgeries for HLHS and HRHS is the Fontan procedure. The Fontan operation was first used in 1968 for the repair of the tricuspid atresia valve and was described by Fontan and Baudet in 1971[7]. Since then it has undergone variations. in the contemporary design of the Fontan operation the superior vena cava is anastomosed directly to the pulmonary artery and, using a conduit that is created inside the baffle, the inferior vena cava is also drained to the pulmonary artery (lateral tunnel technique). The most recent modification of the Fontan operation technique includes the use of an extracardiac interposition graft between the transected inferior vena cava and pulmonary artery[7].

Fetal cardiac intervention consists of in utero techniques which doctors use in patients with congenital heart diseases. There are two categories in which fetal cardiac diseases can fir if it is to be considered for fetal interventions:

- there is a progression in disease process from the time of midgestation diagnosis to birth(diseases that worsen during midgestation and/or late gestation)
- diseases which carry a high risk of death in utero and/or are life threatening at birth and for which fetal cardiac intervention offers a chance of survival[8].

A fetal aortic balloon valvuloplasty is done in HLHS whereas a fetal pulmonary balloon valvuloplasty is done in HRHS. Today , fetal aortic balloon valvuloplasty is performed percutaneously using ultrasound. Fetal aortic valvuloplasty is performed using maternal sedation and epidural anaesthesia. Intramuscular fetal anaesthesia is used to facilitate fetal pain and muscle relaxant is used for appropriate fetal positioning. Once the fetal body is in the proper position, a percutaneous 18 or 19 gauge needle is

inserted into the fetal chest and through the LV myocardial wall with the needle bore pointed in the direction of the LV outflow tract. After that, the stenotic aortic valve is passed a wire. A deflated coronary angioplasty balloon is positioned over this wire at the level of the annulus of the aortic valve. the balloon is inflated up to maximal diameter of ~3.3 mm when the balloon position is confirmed. A procedure is considered technically successful if there is improved anterograde flow across the aortic valve and/or new aortic regurgitation[8].

Fetal pulmonary valvuloplasty done during 21-32 weeks is also done in a similar way but the needle punctures the Right Ventricle. If the Pulmonary Valve was perforated and/or passed and dilated with a balloon catheter, the procedure is considered successful . When The Pulmonary Valve is perforated and passed, but the valve is not dilated with the full diameter of the balloon it is called partially successful procedure [9].

The aim of this survey was to find out more about the staged reconstruction surgeries, at what ages they are done and whether the option for fetal intervention was offered to them.The objective of this study was to find out how many surgeries and what kind of surgeries people with HLHS or HRHS had undergone and whether they were offered the option of fetal intervention and to bring awareness among medical professionals and parents of affected children about the surgeries and interventions available.

II. Material And Methods

A survey was conducted among people whose children were given a diagnosis of Hypoplastic right heart syndrome/ Hypoplastic left heart syndrome or whom themselves had either one of the syndromes. The google form questionnaire consisted of 14 questions on the diagnosis, time period of receiving the diagnosis, surgeries performed, option of fetal intervention given. Data on when the staged reconstruction surgeries or any other surgery were done, whether the surgeries failed an whether the person has other syndromes, whether the person has gotten a heart transplant or is in the process of getting one, whether the person affected is dead or alive.

The google form was circulated in Facebook groups and was also send as email to certain people.

Ethics approval was waived off and Participants were informed that the data collected would be used only for a research article.

III. Results

In the survey conducted, 28 responses were recorded. It could not be determined whether the parent of the child had answered or a person affected by either one of the syndrome had answered in all cases as there wasn't a question for collecting such a data.

Fig. 1 shows the response of the participants of which 79%(n=22) gave the form of CHD as HLHS as the answer while the rest 21%(n=6) gave HRHS as the answer.

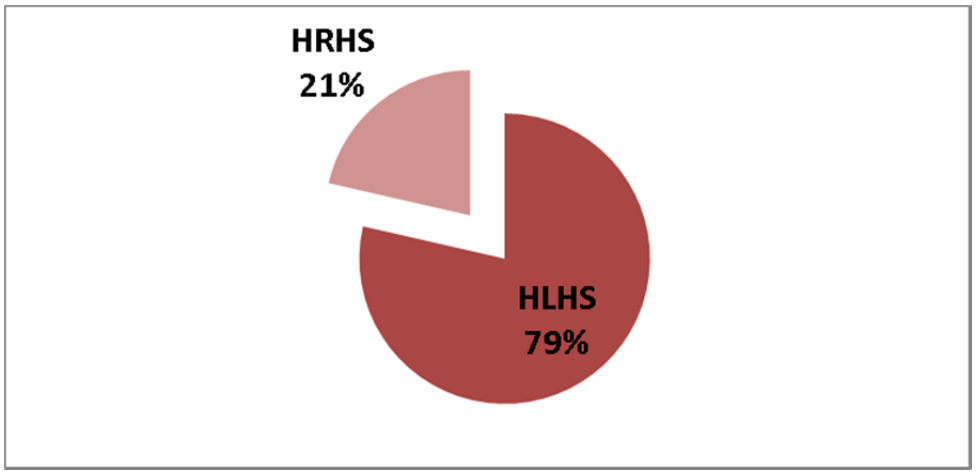

Figure 1- response to the question 'what form of CHD do you/your child have?'

When asked about when the diagnosis was received, 88%(n=23) answered that the diagnosis was received while the mother was still pregnant with the child while 12%(n=5) received the diagnosis after the birth of the baby.

The 3rd question on the questionaire asked the participants about the exact diagnosis of which some answers received were detailed with a number of conditions mentioned while other answers just said HLHS/HRHS. Table 1 shows the answers received for this question. The table shows the exact diagnosis written by the participants except in some cases where HLHS/HRHS was added to make it easy to understand.

Table 1(original table) - response to the question ' What is the exact diagnosis?'

Diagnosis	Number of responses
HLHS	11
HRHS	1
HLHS, Aortic stenosis/Mitral stenosis(AS/MS) and narrowed aortic arch	1
HLHS, Mitral atresia/ Aortic/atresia(MA/AA)	4
HLHS, AS/MS	1
HLHS, MS/AA	1
HLHS , Restricted Atrial septum	1
HLHS, Double outlet right ventricle(DORV),MA/AA	1
HLHS, MA/AA, Mild regurgitation	1
HRHS	1
HLHS, dextrocardia, single ventricle, tricuspid &pulmonary atresia, atrial septal defect, ventricular septal defect	1
HRHS, tricuspid atresia, aortic coarctation, atrial septal defect, ventricular septal defect	1
HRHS, atrioventricular septal defect, pulmonary stenosis, malposed great vessels	1
HRHS, double inlet left ventricle(DILV), Transposition of great arteries, hypoplastic arch, coarctation of aorta, Patent ductus arteriosus(PDA), Patent foramen ovale(PFO), Ventricular septal defect, atrial septal defect, bilateral superior vena cava	1
HRHS, pulmonary atresia with intact septum	1

The 4th question asked whether the option for fetal intervention was suggested to them in which 89%(n=25) participants said that they did not received the option of fetal intervention while 11%(n=3) said that they did receive the option.

Question 5 was ' If you were given the option for fetal interventions , did you do it' to which some answers received give no definite conclusion as to whether the participants understood the question. The responses received are 88%(n=23) say that fetal intervention was not done while 12% (n=5) say that fetal intervention was done. In the previous question only 11% participants received the option for fetal intervention. What this may mean is that either the participants didn't understand the question or maybe they did not get the option for fetal diagnosis from their doctor but had to go around looking for any treatment option for the fetus.

Fig. 2 shows the responses for the question asking about the number of surgeries and it shows a wide range of numbers and in the next question participants were asked whether all surgeries were related for HLHS/HRHS and only 71%(n=20) participants state that all the surgeries were related to HLHS/HRHS while 29% (n= 8) say that all surgeries where not related to HLHS/HRHS. The following question would reveal more about the surgeries done.

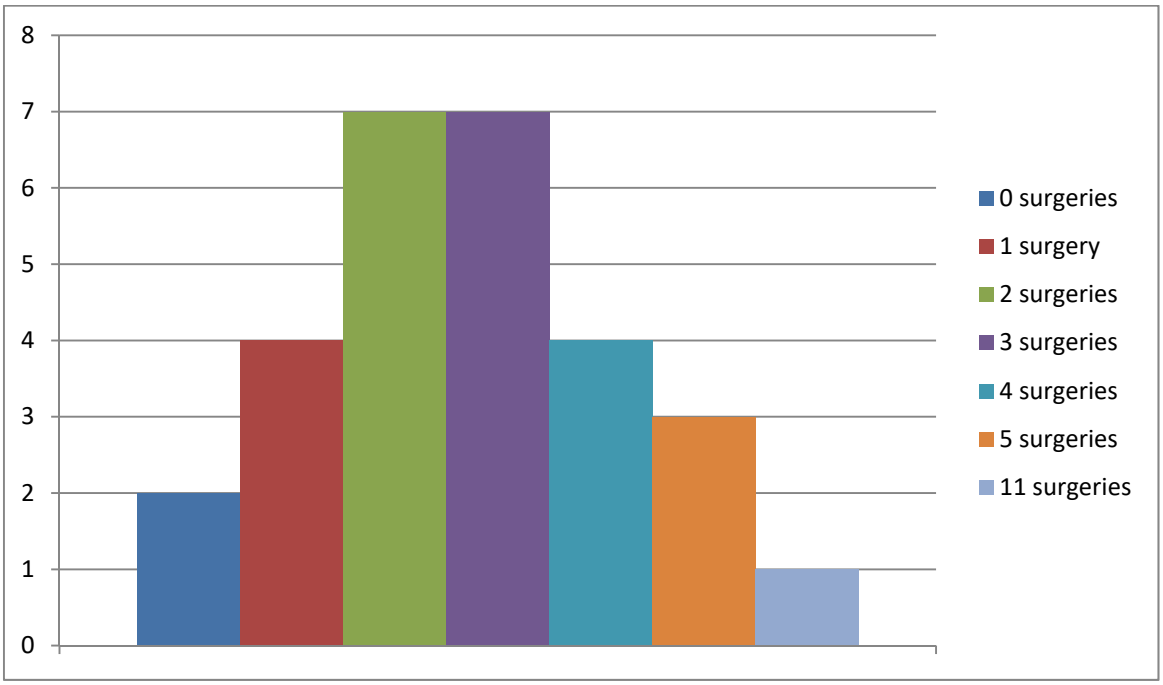

Figure2-response for the question 'how many surgeries have you or your child have had so far?'

Table 2 gives more details about the surgeries underwent it consists of the exact answers given except in some places where the words have been altered to correct spelling mistakes and to group together the answers. From this table we can see that only 1 participant mentions fetal cardiac intervention.

Table 2(original table)– response to the question ' What surgeries have you or your child had so far?

Surgeries		Responses	
None		2	
Norwood		4	
Norwood & Glenn procedures		5	
Norwood, Glenn, Fontan procedures		7	
Fenestrated Fontan(specified)	Just specified as Fontan	1	6
Norwood(BT Shunt),Fontan procedures		1	
Norwood, Nissen fundoplication and G-tube placement, Glenn, Fontan, correction of Right outflow tract obstruction		1	

Fetal heart intervention, balloon angioplasty, open heart surgery, craniotomy with stroke removal	1
Atrial switch and arch repair, Norwood, catheter stent insertion, cardiac shunt, gastrostomy	1
Norwood, Glenn,2 diaphragm plications, G-tube placement	1
Norwood, Glenn, G- tube placement, circumcision	1
Double transplant, Fontan, others	1
Norwood, Glenn, G-tube placement	1
Placement of G- tube	1
Hybrid comprehensive stage 2	1

The next question asked about at what ages did the people undergo the staged cardiac reconstruction surgeries the results of which can be seen in Figures 3a, 3b and 3c.

FIG. 3a shows that from the total of 28 responses, 88%(n=24) responses mentioned undergoing the Norwood procedure of which 75% (n=18) children underwent the surgery aged 1 week or lesser, 8%(n=2) children underwent the surgery aged 2weeks or lesser, 13%(n=3) underwent the surgery aged 3 weeks or under and4%(n=1)underwent the surgery at 17 years old. The participant who had Norwood done at the age of 17 specifically had the BT shunt at that age.

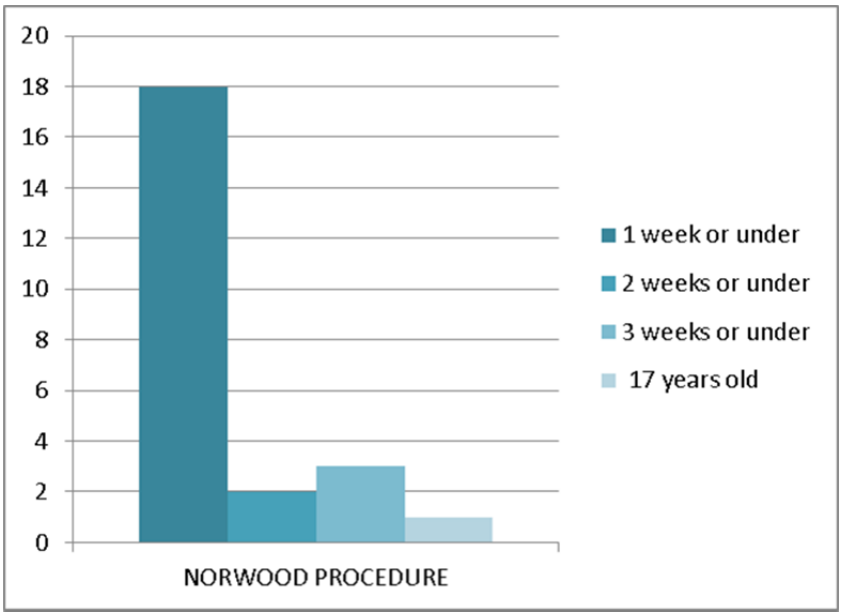

Figure 3a-Ages at which Norwood procedure was done

Fig. 3b shows the 19 responses recorded for having undergone the Glenn procedure , of which 5%(n=1)underwent the procedure at lesser than 3 months old,42%(n=8)of the kids underwent the procedure at 3-4 months of age,21%(n=4)of the kids underwent the procedure at 4-5 months of age, 16%(n=3) of the kids underwent the procedure at 5-6 months of age,11%(n=2) of the children underwent the procedure at 7 months old and 5%(n=1) underwent the procedure at 9 months old.

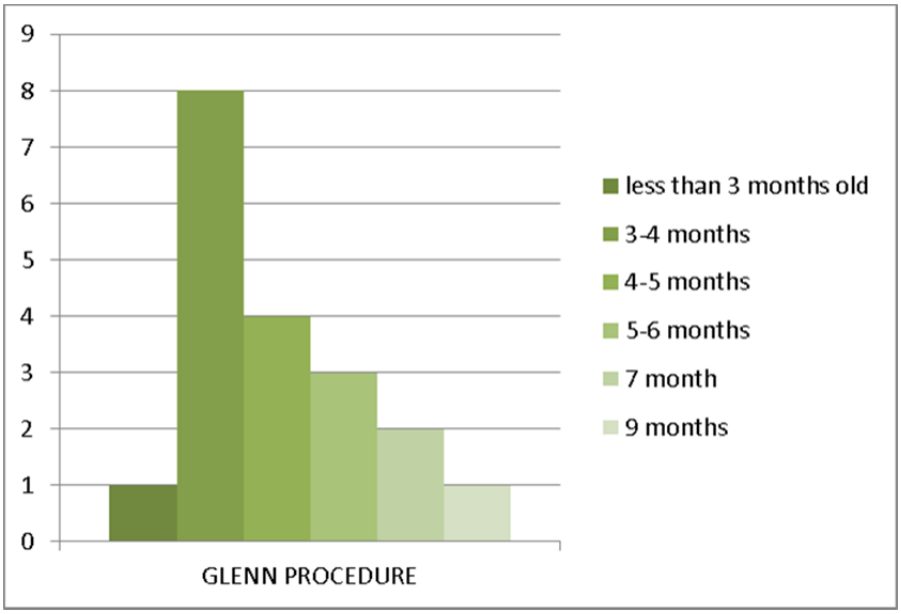

Figure 3b-Ages at which Glenn procedure was done

Fig. 3c show the 9 participants whose responses were recorded as having undergone the Fontan procedure, of which 45%(n=4)underwent the procedure at 2-3 years of age,11%(n=1) of the kids underwent the procedure at 3-4 years of age, 33%(n=3) of the kids underwent the procedure at 4-5 years of age and 11%(n=1) underwent the procedure at 26 years of age.

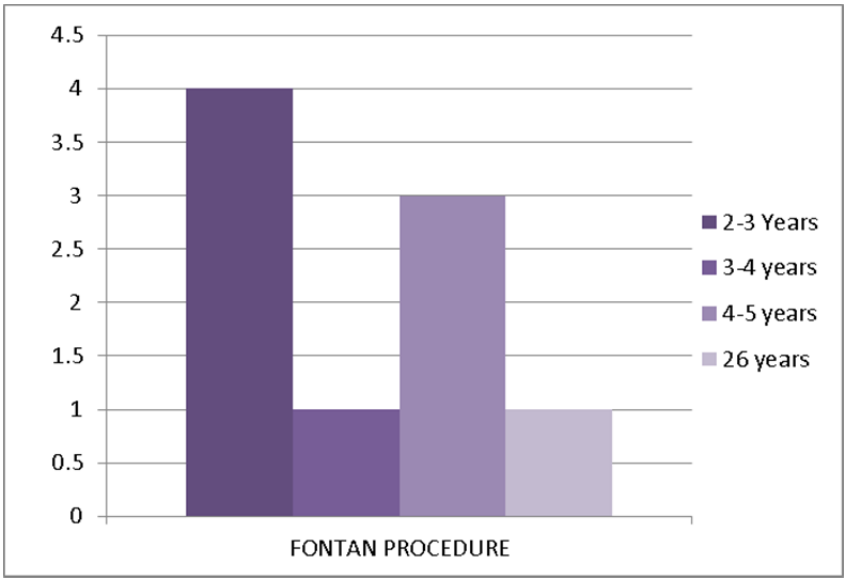

Figure 3c- Ages at which Fontan was done

One of the response was discarded as it just mentioned '9' without specifying days/weeks/months.

3 of the responses mentioned not having undergone any of the staged reconstruction surgeries.

The timelines mention the data which is over the lower limit and equal to or under the upper limit. For example in 3-4 months of age, kids who were over 3 months of age but not 3 months of age and equal to or lesser than 4 months of age were included.

Question 10 asked the participants if any procedures had failed the results. Out of the 26 participants who responded that surgery/surgeries were performed 88% (n=23) responded that the procedures hadn't failed. While 12%(n=3) responded that there was failure of the procedure/procedures.

As the patients may also have other health conditions along with HLHS/HRHS the participants were asked whether they had any other syndromes to which 86%(n=24) said no while 14%(n=4) said yes in which 4(n=1) responded as craniosynostosis , 7%(n=2) responded with hypothyroidism and 3%(n=1) responded with IBS, PCOS. Now hypothyroidism is not a syndrome but the participant may have been unaware of this. Craniosynostosis is also not a syndrome but it is associated with other syndromes. The results are shown in Fig. 4.

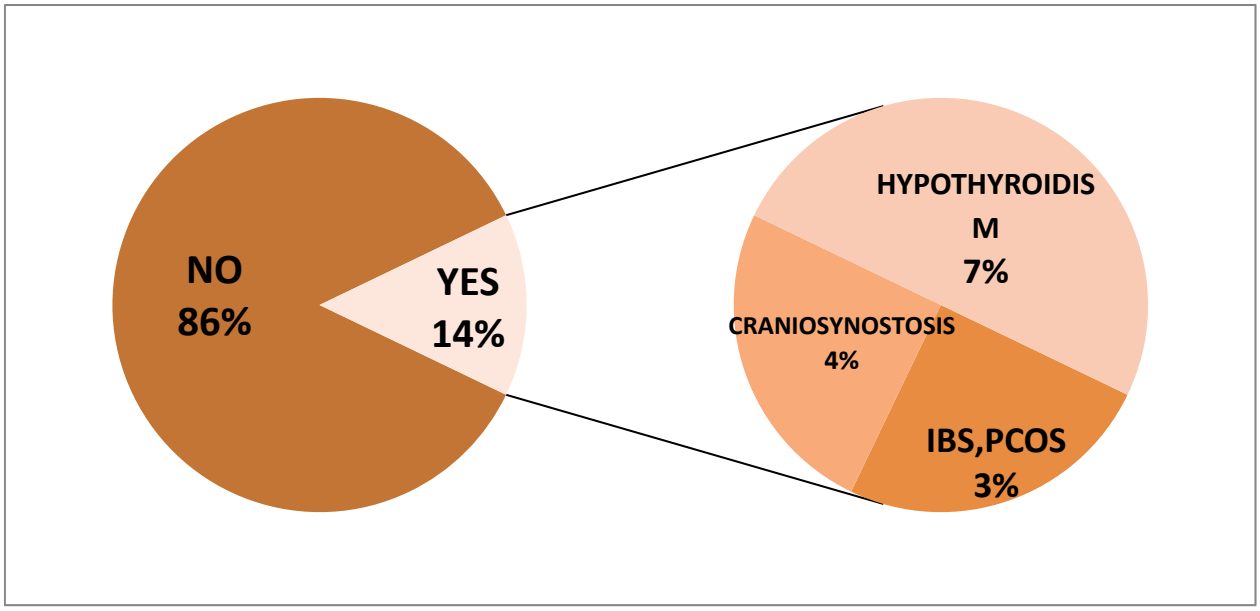

Figure 4- response to the question ‘ Do you or your child have any other syndromes’

Patients with HLHS/HRHS may require a heart transplant. So the participants were asked ‘Did you or your child receive or are in a process of getting a heart transplant’ to which 27 replied no and 1 participant replied as yes.

The next question inquired about whether the person affected with HLHS/HRHS is alive or dead to which 26 participants replied that the person was alive and 2 of them stated that the child due to HLHS.

One of the participant stated that the doctors suggested him not to do the surgery and he was informed that the success rate for the staged reconstruction surgeries are low.

IV. Discussion

This survey was conducted to get to know how many surgeries a person with HLHS/HRHS undergoes and it can be seen that the number varies widely and is also dependent on the fact that the person may have had surgery for some other disorders or diseases. Also we can see that only 3 were given the option of fetal interventions. One response actually states that fetal intervention was experimental at that time and was actually considered for the child but the child wasn’t a candidate. It is worth mentioning that all fetuses aren’t considered candidates for fetal interventions and there is actually a risk of the fetus dying during the procedure. Doctors don’t give that option if they think that the procedure can be done after the birth of the baby. For one participant after consulting with various doctors he was informed that the 3 surgeries do not have a high success rate and the doctors suggested they don’t do them. The child later died due to HLHS after being alive for less than a year.

The Norwood procedure is performed in the neonate stage[10]. In the results shown above we can see that most of the Norwood procedures performed were done in this stage. Age older than 20 days seems to be an independent risk factor for early postoperative mortality and fatal events after the Norwood procedure[11].

The age in which Glenn procedure is done varies in the different papers published. In our survey we can see the youngest patient had it at the age of 2.5 months(< 3months) while the oldest patient was 9 months old when the procedure was performed.

A study showed that though Fontan is usually performed below the age of 2 in the US, the optimum age for the surgery is 3 years[12]. In our survey we see that a lot of the Fontan procedures were done when the patients were 2-3 years of age.

Though there are risks involved in the cardiac reconstruction surgeries doctors are now looking at fetal interventions trying to open the closed valves while the fetus is still inside the mother. Fetal interventions come with its own set of risks but if successful the heart can develop fully though the child may require other surgeries later.

V. Conclusions

Cardiac surgeries and fetal interventions have their own risks and doctors should be aware of these so that they can inform their patients about the options available and the risk factors associated with them. It should be noted that some doctors may not be aware of the success rates of such interventions and hence may not recommend to their patients.

In this study we can see that a lot of the patients had the surgeries performed in the time period recommended by doctors while others either had it earlier or later.

As far as fetal intervention is considered which is still unpopular in some parts of the world and may even be unknown in some places, our study consisted only of 1 patient who had fetal intervention performed, though it was offered to others.

VI. List Of Abreviations

AA- Aortic Atresia

AS- Aortic Stenosis

DILV - Double Inlet Left Ventricle

DORV - Double Outlet Right Ventricle

Hypoplastic left heart syndrome (HLHS)

Hypoplastic right heart syndrome(HRHS)

MA – Mitral Atresia

MS – Mitral Stenosis

PDA - Patent Ductus Arteriosus

PFO – Patent Foramen Ovale

VII. Acklowledgements

Ethics approval and consent to participate – Ethical approval waived off and consent was taken

VIII. Availability of data and material

Data generated or analyzed during this study are included in this published article and can be made available upon request.

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