International Journal of Early Childhood Special Education (INT-JECSE)
DOI:10.9756/INTJECSE/V14I5.539 ISSN: 1308-5581 Vol 14, Issue 05 2022

Case Report on Management and Outcome of Congenital Tricuspid Atresia with Pulmonary Atresia With An Operated Case Of Bidirectional Glenn.

Tanu Prakash Walke[1], Bhagyashree Ganeshpure[2],Shivendra Singh[3], Ashwini Potdukhe[4]

1] GNM, 3rd year, Florence Nightingale College of Nursing,Sawangi (m), Wardha, India, Email: tanuwalke28@gmail.com, 8329072130
2] Nursing Tutor,Florence Nightingale College of Nursing, Sawangi (M) Wardha, India, Email: bhagyashree1706@Gmail.Com, 8805297654
3] Research Consultant, Department of Research and Development, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences, Wardha, Email: shivendra3497@gmail.com
4] Department of Medical-Surgical Nursing, Smt. Radhikabai Meghe Memorial College of Nursing, Datta Meghe Institute of Medical Sciences, Sawangi, Wardha, Maharashtra.

Abstract:
Kreysig (1817) was the first to describe tricuspid atresia, a rare cyanotic congenital cardiac condition with a frequency of 1% to 5%. Without surgical intervention, it is incompatible with extended life. Glenn's anastomosis of the superior vena cava to the right pulmonary artery was effective. Fontan effectively divides the right and left cardiac circulations; this is the preferred surgical technique, with a death rate of seventeen per cent through the age of 20. Rather than the heart pumping blood to the lungs first, the bidirectional Glenn surgery transfers blood directly from the upper body veins to the lungs.This case reports we discussed the management and outcome of congenital tricuspid atresia with pulmonary atresia with an operated case of bidirectional Glenn.
A 3-month-old male child admitted to CVTS department with the major complaint of dyspnoea, cough, and cold since 1month. He has since been transferred to our tertiary care rural hospital, Wardha for further surgical procedures. After routing and physical examination, a doctor declared a tricuspid atresia case for that medication inj. piptaz 400 mg IV TDS, Tab. Lanzol 5 mg OD, tab. Aspirin 75mg 1 tab. In 3ml NS, give 1 ml OD, tab calcimax 2.5 ml twice a day, Syrup. furoped 0.5 ml twice a day, Syrup Ibugesic plus 2.5 ml twice a day, Drop multivitamin 2.5ml od, Syp Zinconia 2.5ml Od, Medium Chain Triglyceride 0.5ml Each Feed, Tab Sildenafil 25_1tab+5ml And Give 1.3ml, Syp Augmentin Dds 2.5ml Bd, Syp Kesol 1ml Tds, Entrogermina Oral Susp. ½ Bd and nebulisation given. After surgical treatment, the child's condition improved.

Keywords: Tricuspid Atresia, Bidirectional Glenn Procedure, Pulmonary Atresia.

Introduction:

Tricuspid Atresia isan uncommoncongenital cyanotic heart anomaly, initially identified by Kreysig in 1817 and classified into three kinds by Kuhne in 1906. [1]It is a congenital heart condition in which a tricuspid valve between two heart chambers does not form. It affects one to five per cent of the population and makes long-term life impossible without surgery. 2 The Glenn in 1958 was the first surgery to successfully use a superior vena cava to right pulmonary artery anastomosis, now known as the bidirectional Glenn anastomosis. [3]
The bidirectional Glenn shunt, also known as a bidirectional cavopulmonary anastomosis, is a surgical procedure used in paediatric cardiac surgery to increase blood oxygenation for individuals with a single functional ventricle because of a congenital heart defect.[4] To treat Single Ventricle Physiology effectively, this approach is required.[5]the purpose of this technique is ventricular deloading, which has been linked to beneficial changes in the SV, such as decreased ventricular end-diastolic volume and suitable ventricular wall thickness geometrical modifications. [6]The absence or agenesis of the tricuspid valve at birth is known as tricuspid atresia. The two most prevalent cyanotic congenital cardiac abnormalities are great arteries and TOF transposition. Tricuspid atresia is the most common cause of cyanosis in people with left ventricular hypertrophy. [8]Tricuspid atresia is a congenital heart defect that affects two out of every 10,000 babies born alive. It is relatively uncommon; accounting for about 1 to 2% of all congenital heart disease occurrences.[9]Tricuspid atresia is the 3rd most frequent cyanotic congenital cardiac defect, with a 90 per cent mortality rate before the age of ten. Although surgical intervention has lowered mortality, the mortality rate remains high due to unfavourable anatomy.[10]Patients' average life expectancy has increased to 35 to 40 years, compared to 70 to 80 years for the general population. [11]

Congenital cardiac illness affects about 1 per cent of all live births, with 50 per cent of those babies being girls.[12]Heart surgery and post-surgical treatment have advanced to the point where there is now a CHD that affects a substantial number of women, most of whom wish to start a family. In these women, pregnancy is connected to cardiac difficulties, which occur in about 10% of pregnancies.[13]With tricuspid atresia, a baby, child, or adult cannot get enough oxygen through their bodies. [14] This condition causes people to tyre easily, be short of breath, and have blue-tinged skin. Multiple surgeries are used to treat it. Most babies with surgery for tricuspid atresia live into adulthood, though follow-up procedures are frequently required. [15]

Patient information:
The three-month-old child was admitted to the CVTS ward with the chief complaints of breathlessness, cough and cold since 1month. The child wascase of congenital tricuspid atresia with pulmonary atresia now his brought to tertiary care hospital for the further surgical procedure of that.

Primary Concern and symptoms of patient:
Patient well before 1month after that patient starting breathlessness recurrent cough cold so after that patient seen near private doctor.it advice 2D echo. patient diagnosed with tricuspid atresia with pulmonary atresia so patient advice surgical management. For that patient brought here to Tertiary care hospital, Wardha in CVTS ward. Later onmanage of that condition bidirectional Glenn surgery was done.

Medical family and psychosocial history: For the same complaint seen near private doctor Previous Treatment Feronia XT Drop 0.5ML OD, Tab Ciplar 10mg 1/4th .there was no other medical history. He was diagnosed at birth with tricuspid atresia. He belongs to a nuclear family. There are four members in their family. All family members are healthy except the Patient and maintain good interpersonal relationships. The child's father occupation was a worker, and his socioeconomic status is good. No psychological history of the child.

Birth history- full-term normal delivery of a child; birth weight was 2.5 kg .vaccination is done till 2 ½ months.

Physical examination and clinical findings: on admission, the physical examination showed the patient was conscious, cooperative, and well-oriented. His body build was moderate, and his personal hygiene was maintained properly. His weight was 6.2 kg and his height was 60.2 cm, pulse was 120/min, his respiration was 30/min, his temperature was afebrile, and his general examination was moderate. His nutrition status was poor. Facial expressions were dull, irritable, and severe cry present. The abdominal examination was soft.

Timeline: As narrated by the mother,thechild was diagnosed a known case of tricuspid atresia present since birth. Now a child is brought to tertiary care hospital, Wardha for further management. Hereafter all investigation of blood testing, urine testing, LFT, and KFT need for operation was done. Later on, her Bidirectional Glenn surgery was done. After the operation patient shifted to ICU; during the hospital, stay patient was on the following medicationinj. piptaz 400 mg IV TDS, Tab. Lanzol 5 mg OD, tab. Aspirin 75mg 1 tab. In 3ml NS, give 1 ml OD, tab calcimax 2.5 ml twice a day, Syrup. furoped 0.5 ml twice a day, Syrup Ibugesic plus 2.5 ml twice a day, Drop multivitamin 2.5ml od, Syp Zinconia 2.5ml Od, Medium Chain Triglyceride 0.5ml Each Feed, Tab Sildenafil 25_1tab+5ml And Give 1.3ml, Syp. Augumentin Tds 2.5ml Bd, Syp Kesol 1ml Tds, Entrogermina Oral Susp. ½ Bd and nebulisation given with NS, a child was stable.

Diagnostic assessment: On the basis of patient history, physical examination and Routinginvestigation was done such as blood Total RBC Count8.11, urine test ,Coagulation profile was normal, micro report urine5-10 RBC/HPF, No pus cell, No organism seen, virological report all non-reactive,Body Fluid (Pleural), RBS was done.

Diagnostic challenges: No challenges during diagnostic evaluation.

Diagnosis: Known case of tricuspid atresia with operated case bidirectional Glenn surgery was done.

Prognosis- Patient's prognosis was satisfactory.

Therapeutic interventions:
In this case of tricuspid atresia, treated medical treatment and nutritional supplements. That is Enterogermina Oral Susp. ½ Bd, Syp Ibugesic Plus 2.5ml sos, Vitanova D3 Sachet 6000 IU Once In A Week , Drop Multivitamin 2.5ml Od , Syp Furoped 0.5ml Bd, Syp Calcimax -P 2.5ml Bd, Tab Aspirin 75mg 1tab+3ml Ns And Give 1ml OD.was given to the patient as per the doctor's order.The nursing perspective is that IV fluids were provided to maintain fluids and electrolyte imbalances. Check vital signs and blood pressure every hour. Maintain an hourly intake and outflow chart. Daily wound cleaning and dressing.

Follow-up and outcomes: Thechild's condition wasimproved. Advicedthe mother for the daily cleaning and dressing of wound. Avoid high cholesterol diet,eat a healthy balanced diet, regular health check-ups, maintain personal hygiene and sanitation, and take proper medication by doctor's order.

Discussion:

Patient was well before 1month after that patient starting breathlessness recurrent cough cold so after that patient seen near private doctor it advice 2d echo patient diagnosed with tricuspid atresia with pulmonary atresia so patient advice surgical management (BDG). For that patient admitted in CVTS ward. All routine investigation done. Patient operated bidirectional Glenn surgery was done. AfterOT patient shifted ICU during hospital stay patient on above medication**.**Patient now stable and discharge in stable condition**.**

A child with tricuspid atresia who had a rare congenital heart defect that made long-term survival impossible without cardiac surgery was able to survive and function at NYHA I–II for three months with only the Glen procedure, without any of the ventricular, dysrhythmic, systemic, or organ dysfunction associated with the Fontan circulation.[16] after Fontan, patients saturations varied from 90% to 94%. However, the index case had a steady-state saturation of 74%, demonstrating that great cognitive function is not compromised. This indicates that the Glenn procedure is merely an alternative to the Fontan for certain anatomical types of tricuspid atresia, with no need to proceed to the Fontan, which has well-documented drawbacks such as additional cardiac surgery and associated perioperative and intraoperative morbidity and mortality. [17]

This is the first case report of tricuspid atresia three months after Glen alone, including normal superior vena cava flow towards the right pulmonary artery, healthy pulmonary blood flow, and no signs of pulmonary hypertension or other Fontan circulatory dysfunctions.[18-23]

To prevent the ductus from shutting after birth, an infant with tricuspid atresia may require medication. It may be necessary to perform a technique or surgery to create an opening between the atria or to link the aorta to the pulmonary artery. A hole between the ventricles is present in tricuspid atresia affects many babies (ventricular septal defect).[14-26]

When performed following a systemic pulmonary arterial shunt, the Glenn anastomosis was a success.Children underwent the Fontan procedure (hospital mortality 16.6 per cent). After the third month following surgery, no late fatalities have occurred. Despite the fact that the typical follow-up for this surgery is only two years, 88% of surviving and go on to enjoy normal lives, with two-thirds of them receiving no treatment. [27-28]

Informed consent: His parents were informed before taking this case**.**

Conclusion:

Tricuspid atresia is a difficult CHD with a significant death and morbidity rate.In this case we discussed the case of tricuspid atresia with pulmonary atresia and its surgical management and its outcomes. After surgical procedure patient condition was stable and prognosis was satisfactory.

Conflict of interest: Noconflict of interest
Funding: None

References:

1. WILLIAMS-PHILLIPS, S. Tricuspid Atresia 18 Years Post Glenn: Is Fontan Necessary in All Cases? The West Indian Medical Journal, 2015, 64.3: 294.
2. MOND, Harry G.; KARPAWICH, Peter P. Pacing options in the adult patient with congenital heart disease. John Wiley & Sons, 2008.
3. SALIK, Irim; MEHTA, Bhupen; AMBATI, Shashikanth. Bidirectional Glenn Procedure or Hemi-Fontan. 2020.
4. ANDERSON, Jeffrey B., et al. Lower weight-for-age z score adversely affects hospital length of stay after the bidirectional Glenn procedure in 100 infants with a single ventricle. The Journal of thoracic and cardiovascular surgery, 2009, 138.2: 397-404. e1.
5. WALKER, Scott G.; STUTH, Eckehard A. Single-ventricle physiology: perioperative implications. In: Seminars in pediatric surgery. WB Saunders, 2004. p. 188-202.
6. MOHAMED, Marwa, et al. A Rare Case of Successive Pregnancies in a Woman with Complex Cyanotic Congenital Heart Disease after Glenn Palliation. World Journal of Cardiovascular Diseases, 2017, 7.12: 423-428.
7. GOLDSTEIN, Andrew H., et al. Partial support with a centrifugal left ventricular assist device reduces myocardial oxygen consumption in chronic, ischemic heart failure. Journal of cardiac failure, 2005, 11.2: 142-151.
8. RAO, P. Syamasundar. Management of congenital heart disease: state of the art—part II—cyanotic heart defects. Children, 2019, 6.4: 54.

9. HOFFMAN, JULIEN IE. Natural history of congenital heart disease: problems in its assessment with special reference to ventricular septal defects. Circulation, 1968, 37.1: 97-125.
10. KEATING, P.; VAN DER MERWE, P. L.; SHIPTON, S. Tricuspid atresia-profile and outcome. Cardiovascular Journal of Africa, 2001, 12.4: 202-205.
11. SUMAL, Anoop S.; KYRIACOU, Harry; MOSTAFA, Ahmed MHAM. Tricuspid atresia: Where are we now?. Journal of Cardiac Surgery, 2020, 35.7: 1609-1617.
12. WHITTEMORE, Ruth; HOBBINS, John C.; ENGLE, Mary Allen. Pregnancy and its outcome in women with and without surgical treatment of congenital heart disease. The American journal of cardiology, 1982, 50.3: 641-651..
13. GONGORA, Maria Carolina; WENGER, Nanette K. Cardiovascular complications of pregnancy. International journal of molecular sciences, 2015, 16.10: 23905-23928.7.
14. RAO, P. Syamasundar. Single Ventricle—A Comprehensive Review. Children, 2021, 8.6: 441.
15. FROCK, Brittany Watson; JNAH, Amy J.; NEWBERRY, Desi M. Living with tricuspid atresia: case report with review of literature. Neonatal Network, 2017, 36.4: 218-228.
16. TRAN, Derek, et al. Recommendations for exercise in adolescents and adults with congenital heart disease. Progress in Cardiovascular Diseases, 2020, 63.3: 350-366.
17. HEYDARIAN, Haleh C.; MADSEN, Nicolas L.; MARINO, Bradley S. Long-term outcomes in congenital heart disease. In: Pediatric Critical Care Medicine. Springer, London, 2014. p. 417-440.
18. YAMAUCHI, TOYOSHIGE; CAYLER, GLEN G. Ebstein's anomaly in the neonate: A clinical study of three cases observed from birth through infancy. American Journal of Diseases of Children, 1964, 107.2: 165-172.
19. ZENG, Zhandong, et al. Current diagnosis and treatments for critical congenital heart defects. Experimental and therapeutic medicine, 2016, 11.5: 1550-1554.
20. DE BRUX, Jean-Louis, et al. Tricuspid atresia: results of treatment in 115 children. The Journal of thoracic and cardiovascular surgery, 1983, 85.3: 440-446.
21. Ghule, Aishwarya, Shilpa Abhay Gaidhane, Sourya Acharya, Charan Bagga, and Anil Wanjari. "Libman-Sacks Endocarditis Involving Tricuspid Valve in Systemic Lupus Erythematosus - A Case Report." Journal of Evolution of Medical and Dental Sciences 10, no. 16 (April 19, 2021): 1179–81. https://doi.org/10.14260/jemds/2021/251.
22. Acharya, S., Ghewade, B., Shukla, S., Prothasis, M., 2020b. Electric Shock-Induced Pulmonary Hemorrhage - A Rare Phenomenon. INDIAN JOURNAL OF RESPIRATORY CARE 9, 127–128. https://doi.org/10.4103/ijrc.ijrc_49_19
23. Bawiskar, N., Dronamraju, S., Godhiwala, P., Rathi, C., Kumar, S., 2020b. Pickwickian syndrome with severe pulmonary hypertension: Deciphering the early connection. MEDICAL SCIENCE 24, 3679–3682.
24. Gandhasiri, D., V., Dhamgaye, T.M., Jadhav, U., Ghewade, B., 2020. A Case of Disseminated Extensively Drug Resistant Extrapulmonary Tuberculosis. JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH 14, LD1–LD3. https://doi.org/10.7860/JCDR/2020/44916.14183
25. Gupta, K.R., Sawarkar, G., Jadhav, U., 2020. Critical appraisal of pipasa in chronic obstructive pulmonary disease. INTERNATIONAL JOURNAL OF AYURVEDIC MEDICINE 11, 44–49.
26. Gupte, M., Jadhav, U., Ghewade, B., Sherekar, D., Gandhasiri, D., 2020. To study the incidence of lung cancer in patients of newly and previously diagnosed Chronic Obstructive Pulmonary Disease at AVBRH. MEDICAL SCIENCE 24, 658–663.
27. Lahole, S., Acharya, S., Andhale, A., Takey, P., 2020a. Acute Pulmonary Embolism (PE) with Acute Cor-Pulmonale treated successfully by Catheter Directed Fibrinolysis (CDF). MEDICAL SCIENCE 24, 2727–2731.
28. Singh, R.K., Singhania, A., Mishra, G.V., Patwa, P.A., 2020a. Congenital Pulmonary Airway Malformation in One of the Monochorionic Diamniotic Twins - A Rare Encounter. JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES-JEMDS 9, 3694–3696. https://doi.org/10.14260/jemds/2020/810