

# DOWNLOAD PDF ANAESTHETIC MANAGEMENT OF TETRALOGY OF FALLOT

## Chapter 1 : considerations | Tetralogy of Fallot Considerations

*Tetralogy of Fallot is the most common form of cyanotic heart disease and accounts for % of all cases of congenital heart disease ( live births in the UK). The cause of TOF is unknown, but.*

Its hallmark anterior and superior infundibular septal displacement gives rise to the tetrad of ventricular septal defect, aortic override, infundibular obstruction, and right ventricular RV hypertrophy<sup>2</sup>. Any disease complicated by severe maternal hypoxemia is likely to lead to miscarriage, poor fetal growth, preterm delivery or fetal death. There is a relationship between chronic hypoxemia and the polycythemia it causes with the outcome of pregnancy. With satisfactory surgical correction prior to pregnancy, maternal risks are decreased dramatically, and fetal environment is improved. Intracardiac repair has permitted survival into the childbearing years and excellent quality of life<sup>6</sup>. Long-term complications usually relate to functional competence of the RV outflow tract and its secondary effects on ventricular and atrial myocardial function. Pulmonary regurgitation or stenosis may result in RV dysfunction and failure, progressive tricuspid valve regurgitation, atrial and ventricular arrhythmias, and sudden cardiac death<sup>7</sup>. Case Report A year-old unbooked female primigravida was posted for emergency cesarean section during labour. She had complaints of dyspnoea, early fatigability and history of cyanotic spells during heavy exertion before her pregnancy. She had history of taking beta blocker propranolol and digitalis off on. Her cesarean section was decided because of failed progression of labor due to cephalo-pelvic disproportion. Clubbing and murmur was present. As patient came in emergency operating room, her other investigations were not available at time of surgery. General anaesthesia was planned for her surgery. Ranitidine and Metaclopromide was given half an hour before surgery. Amoxicillin was also given for prophylaxis. Uterus contracted slowly spontaneously. On completion of surgery the patient was reversed with neostigmine 0. She was further referred to cardiothoracic department. Discussion Maternal heart disease complicates 0. The complex of anatomic malformation results from an anterior displacement of the conoseptum toward the right ventricle creating a malalignment VSD and a narrowing of the outflow tract of the right ventricle RV 2. The aorta is displaced anteriorly, straddling the muscular septum and arising from both ventricles. The obstruction to outflow of the RV usually involves the infundibulum of the RV but can arise from the pulmonary valve, its annulus, the main pulmonary artery or even in the peripheral pulmonary arteries. Elevated pressures in the RV from outflow obstruction and exposure to systemic pressure from overriding aorta lead to compensatory RV hypertrophy. The main characteristic of TOF is cyanosis. Cyanosis can result from three separate Mechanisms. Inadequate pulmonary blood flow, right to left shunting or intrinsic pulmonary disease. In TOF, cyanosis results from a right-to-left shunt at the level of ventricles and inadequate pulmonary blood flow. Because of the outflow obstruction, blood ejected from RV crosses the VSD and enters the overriding aorta. This reduces the amount of pulmonary blood flow available for oxygenation and adds desaturated blood to the systemic circulation. Pressures in the right ventricle are near to the systemic pressure. The likelihood of a favorable outcome for the mother with TOF depends upon the functional cardiac capacity of the patient before pregnancy, other complications that further increase cardiac load, and quality of medical care provided throughout pregnancy and surgical correction of the anomaly before conception. Pregnant mothers with TOF are affected differently depending upon if they remain uncorrected, have palliative or definitive procedure or they have residual defects after these procedures. The principle danger for a pregnant woman with TOF is cardiac decompensation because of inability to meet the additional demands imposed by the physiological changes of pregnancy and parturition. If present, infection, hemorrhage and thrombo-embolism compound the risk. The cardiovascular changes of pregnancy may unmask residual or recurrent TOF in patients with corrective procedures, who have been asymptomatic throughout their life after TOF repair <sup>6</sup>. Before successful intracardiac repair of TOF was introduced in the s, few patients reached childbearing age, and successful pregnancy was uncommon. Pregnancies were characterized by spontaneous abortions, stillbirths, and

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premature deliveries. Chronic hypoxemia in such patients leads to adaptations to provide adequate tissue oxygenation i.e. As no specific technique is suggested for such patients and anaesthesia carries considerable risk, the management should be based on avoiding changes that would increase the magnitude of R to L shunt, dehydration should be avoided to improve the circulating volume by decreasing the blood viscosity. Both general and regional techniques have been employed successfully in parturient with TOF. Regarding cesarean section, general anesthesia GA is probably the technique of choice. GA with endotracheal intubation provides airway protection, eliminates work of breathing and may reduce oxygen consumption. The complications of controlled mechanical ventilation include decreased venous return as well as ventricular dysfunction, compression of pulmonary vessels, hypoxemia, hypo or hypercarbia and acidemia. The choice of anesthetic drugs may not be of prime importance. In patients with dynamic right ventricular outflow obstruction, increases in heart rate and contractility should be avoided as they will worsen the obstruction and cyanosis. Anesthetic drugs and adjuncts having vagolytic or sympathomimetic effects should preferably be avoided. Epidural catheter techniques offer continuous, titrated anesthesia or analgesia. In this case we used general anaesthesia as patient came as emergency with known TOF but without taking any medicine regularly and with cardiac decompensation. She had not gone any palliative or corrective procedure. To achieve this ketamine was used for induction and phenylephrine to increase SVR. She was preloaded with ml RL before induction. Patient was hyperventilated to maintain slightly alkalosis. Oxytocin was not used to avoid sudden increase in preload. Invasive monitoring CVP and arterial blood pressure could not be monitored in that case because of unavailability. Patient was managed properly. Patients with tetralogy of Fallot with pregnancy need special care by a team consisting of obstetrician, cardiologist and anesthesiologist among others.

## Chapter 2 : Tetralogy of Fallot Rx

*Tetralogy of Fallot (TOF) is a common congenital heart defect in children. Perioperative considerations include preoperative preparation for surgery, intraoperative anesthetic management, and common postoperative issues in the intensive care unit.*

In these individuals, blood shunts completely from the right ventricle to the left where it is pumped only through the aorta. The lungs are perfused via extensive collaterals from the systemic arteries, and sometimes also via the ductus arteriosus. Diagnosis[ edit ] A CXR Chest X-Ray of a child with tetralogy of Fallot Congenital heart defects are now diagnosed with echocardiography , which is quick, involves no radiation, is very specific, and can be done prenatally. Before more sophisticated techniques became available, chest x-ray was the definitive method of diagnosis. The abnormal " coeur-en-sabot " boot-like appearance of a heart with tetralogy of Fallot is classically visible via chest x-ray, although most infants with tetralogy may not show this finding. This allows more blood flow to the lungs by decreasing shunting of deoxygenated blood from the right to left ventricle through the VSD. There are also simple procedures such as squatting and the knee chest position which increase systemic vascular resistance and decrease right-to-left shunting of deoxygenated blood into the systemic circulation. Taussig , and lab assistant Vivien Thomas at Johns Hopkins University developed a palliative surgical procedure, which involved forming a side to end anastomosis between the subclavian artery and the pulmonary artery. This first surgery was depicted in the film *Something the Lord Made*. This redirected a large portion of the partially oxygenated blood leaving the heart for the body into the lungs, increasing flow through the pulmonary circuit, and greatly relieving symptoms in patients. The first Blalock-Thomas-Taussig shunt surgery was performed on month-old Eileen Saxon on November 29, with dramatic results. These are no longer used. Currently, Blalock-Thomas-Taussig shunts are not normally performed on infants with TOF except for severe variants such as TOF with pulmonary atresia pseudotruncus arteriosus. The first total repair of tetralogy of Fallot was done by a team led by C. Walton Lillehei at the University of Minnesota in on an year-old boy. The open-heart surgery is designed to relieve the right ventricular outflow tract stenosis by careful resection of muscle and to repair the VSD with a Gore-Tex patch or a homograft. Additional reparative or reconstructive surgery may be done on patients as required by their particular cardiac anatomy. This progresses to heart failure which begins in the right ventricle and often leads to left heart failure and dilated cardiomyopathy. Mortality rate depends on the severity of the tetralogy of Fallot. Untreated TOF also causes delayed growth and development, including delayed puberty. It is recommended that they follow up at a specialized adult congenital heart disease center.

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## Chapter 3 : Anesthetic management of a parturient with uncorrected tetralogy of Fallot for Cesarean section

*Anesthetic objectives for patient's with Tetralogy of Fallot: maintain intravascular volume and SVR. Avoid increases in PVR as may be caused by acidosis or increased airway pressures. Drugs such as Ketamine are recommended for induction because ketamine increases SVR and does not increase the right to left shunting.*

By the end of the discussion, the learner will be able to: Describe the changes in cardiovascular physiology that occur during pregnancy. Acknowledge the potential late outcomes after TOF repair. Discuss anesthetic considerations for neuraxial anesthesia in a parturient with repaired TOF. This accounts for approximately 1, new cases annually. Occurrence of serious adverse cardiac events during labor in adult patients with a history of remote TOF repair varies from relatively uncommon to more than an appreciable risk. Singh, Bolton, and Oakley concluded from their research that pregnancy in a patient with TOF repair can be managed in a normal manner with no special precautions as it is highly unlikely that there will be any complications. None of the 31 parturients experienced syncopal episodes or chest pain. These papers highlight the need for increased vigilance over this group of women for uncommon but potentially serious cardiac complications, especially ventricular arrhythmia and heart failure. This problem-based learning discussion PBLD is designed to facilitate a group exchange focused upon clinical decision-making when selecting neuraxial anesthetic techniques in an adult parturient with a history of a remotely repaired TOF. A clinical scenario is provided as a means to enable review of TOF and repair techniques and understand the most frequently encountered remote sequelae. After a brief overview of the defect and of common palliative and corrective surgeries for TOF repair, participants proceed to a specific discussion concerning the management of an adult parturient with repaired TOF and neuraxial anesthetic technique options. Questions that accompany the case guide the conversation, and the answers provided are based on a current literature review. The target audience for this PBLD is physician anesthesiologists, certified registered nurse anesthetists, anesthesiology residents, medical students, and other physicians and health professionals who would have involvement in the care of a parturient with a remote TOF repair undergoing a neuraxial anesthetic technique for the management of labor pain. In general, pregnancy outcomes in parturients after a repaired TOF with neuraxial anesthesia are favorable. Additional resources that informed the creation of this discussion are noted in Appendix C. The case stem and associated questions were given to participants 4 days in advance of the group session in order to provide adequate time to review the pertinent topics. Review was guided by, but not limited to, the suggested references provided at the end of this case. Participants were encouraged to examine the relevant medical literature and to then develop their own preliminary thoughts on the management of this case. The case stem with references explained the case that the PBLD would be centered on. The case stem included references; the participants were encouraged to explore the references in order to gain a better understanding of TOF in general. At the start of the PBLD, the facilitator reviewed the case stem with the participants, including the pertinent medical history, vital signs, and results of initial investigations. After ensuring that all of the participants fully understood the clinical scenario, the facilitator began the learning discussion by posing the first discussion question to the participants. The facilitator made sure to impart to the participants that this was a group learning environment and that people should feel free to share their knowledge and ideas. The learning discussion provided with the PBLD helps the facilitator guide the ensuing discussion for each question, as it contains very specific answers to each of the discussion questions. After completing the first nine discussion questions, the facilitator then reviewed the labor course with the participants. After again ensuring that the participants fully understood that portion of the clinical course, the facilitator continued to the last four discussion questions. At the conclusion of the PBLD, the facilitator asked the participants if they felt that all of the educational objectives had been met during the 1-hour discussion. A survey was then distributed to all participants; this allowed them to give anonymous feedback about their experience during the PBLD. Each attendee filled out an anonymous evaluation to help us to improve the quality of the PBLD. The evaluation

consisted of four questions. The questions are listed below: What was your overall impression of the session? Did you learn anything from the session? Will this change your practice in the future? Do you have any ideas for improvements? Prior to the second PBLD, learners were given additional time 2 weeks vs. The list of references given to the learners was shortened to four. Each of the provided references was a website; this was done so that busy learners could review the information on a computer or even a smart phone. The new group of PBLD participants included some faculty members, some of the same anesthesiology residents from the first session, new CA-1 residents, medical students, and some nurse anesthetists. On this occasion, we allotted 90 minutes for the discussion, and we allowed the natural flow of the discussion to dictate the order in which we posed the discussion questions. We made sure to involve each learner in the discussion from the beginning and incorporated many new ideas and potential clinical scenarios into the PBLD. At the conclusion of the second session, each participant was asked to anonymously submit feedback for the following questions: If you found this discussion helpful, please describe what aspects were most useful. Do you have any suggestions for improving the PBLD? Is there anything that you may adopt into your clinical practice as a result of this discussion? Is there any topic that you feel required more in-depth discussion? After receiving additional feedback from editors, we modified the stem significantly to allow for a better flow to the PBLD. We attempted to create a more robust clinical pathway with clinical questions that were placed at crucial clinical times. We then showed this newly designed stem to residents and faculty members who had previously participated in our PBLD sessions and asked for feedback on the changes. Results Five anesthesiology residents, six anesthesiology faculty members, and one fourth-year medical student participated in the initial offering of the PBLD. Two of the faculty members, including the faculty moderator, were cardiothoracic fellowship-trained and had significant experience with congenital heart disease. Although the participants were given 4 days to review the topics and references, many admitted that they had only looked at a few references the night before the PBLD session. The PBLD took 70 minutes to complete. The first portion of the PBLD, the case stem and nine discussion questions, took approximately 40 minutes to complete. The second portion, which included the labor course and four discussion questions, took 30 minutes to complete. Ten of the 12 participants were extremely active in the discussions that took place for each question. An anesthesiology faculty member and the medical student did not actively participate in the discussion and preferred to listen to the ideas that were being reviewed. After completion of the PBLD, all of the participants were given a survey to complete. All 12 responses were positive. Each participant stated that he or she acquired knowledge during the PBLD. All five of the residents and the medical student specifically commented on how they appreciated information on congenital heart disease and its implications in adults after the repair. Four of the participants gave specific suggestions on how the PBLD could be improved. The second offering of the PBLD included four anesthesiology faculty members one had attended the first offering , three CA-2 anesthesiology residents all three had participated in the first offering , four CA-1 residents none had attended first offering , one medical student a fourth-year medical student who had not participated in the initial PBLD , and two nurse anesthetists. The participants had been given 2 weeks to prepare for the PBLD. Although most of the participants stated that they had waited until the previous night to review the documentation, two faculty members and four of the CA-1 residents stated that they had reviewed the information much earlier. The repeat participants also commented that they appreciated the extra time that they had received to review the information. The PBLD took 90 minutes to complete. The first portion of the PBLD, the case stem and eight discussion questions including added clinical paths , took approximately 60 minutes to complete. The second portion, which included the labor course and four discussion questions including added clinical paths , took 30 minutes to complete. All 14 participants in the second offering of the PBLD were extremely active in the discussions that took place for each question. After receiving feedback from the initial PBLD, the facilitators were diligent in ensuring that all learners felt comfortable asking questions and participating in the discussion. After completion of the PBLD, all of the participants were given the revised survey to complete. All 14 responses were favorable. All participants stated that they had found the

discussion to be very helpful. All four of the participants who had also attended the first PBLD commented on how they appreciated the different clinical paths that we took during the second offering of the PBLD. They agreed that it was worth attending the second session to absorb that additional information. All four of the faculty members agreed that they had learned information that would likely change their practice in the future. Ten of the participants stated that they had no suggestions for improving the PBLD. One of the faculty members stated that this could be used as a grand rounds topic to involve more of the anesthesiology department. A CA-1 resident reported that he would like the PBLD to have been shorter, as the length of the discussion made it difficult to keep his attention. None of the learners gave any answers to the question asking if there were any topics that needed more in-depth discussion. After receiving additional feedback from editors and revising the PBLD to create a better clinical flow, we asked two faculty members and the three CA-2 residents who had previously participated in the prior PBLD offerings to give us feedback. We are happy to report that all the feedback received was extremely positive. She represents a growing number of people with repaired congenital heart disease who will now live well into adulthood as a result of profound advances in modern medicine and cardiothoracic surgery. As the operative techniques to treat these diseases improve, anesthesiologists are increasingly likely to encounter adult patients with complex cardiovascular physiology due to repaired congenital heart disease. Design of the PBLD proved to be a difficult process. We felt it was imperative that all learners, including medical students, anesthesia residents, and faculty members, were able to learn a significant amount during the 1-hour session. For that reason, we decided to create basic and advanced educational objectives. We decided that this would ensure that each group, regardless of level of education, would be able to gain knowledge from the PBLD. The first three educational objectives were basic. We felt that the medical students and anesthesia residents would be able to learn the most from those topics. The second three educational objectives were advanced topics. We felt that those topics would likely be more appropriate for the anesthesia residents and faculty. The information that we received from the evaluations was very helpful to us. The fact that all of the attendees stated that they had learned a great deal during the PBLD was particularly important to us because, during the creation of the PBLD, we had struggled over how to ensure that learners of all levels would benefit from the session. We were also excited to find out if the faculty would change their practice as a result of the session; we were ecstatic to discover that two-thirds of the anesthesia faculty members reported that they would change their practice as a result of the PBLD. Finally, we were anxious to learn what improvements our learners would suggest for the PBLD. First, we distributed the PBLD materials 2 weeks in advance to give busy learners adequate time to prepare.

## Chapter 4 : Internet Scientific Publications

*INTRODUCTION. Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, characterised by aortic overriding, right ventricular hypertrophy, pulmonary stenosis (PS) and ventricular septal defect (VSD).*

Pregnancy with Uncorrected Tetralogy Of Fallot: The Internet Journal of Anesthesiology. Abstract Among the congenital heart diseases causing intra cardiac R-L shunts Tetralogy of Fallot is the most common syndrome, characterized by presence of VSD, aortic overriding, pulmonary artery outflow obstruction and right ventricular hypertrophy. If remains uncorrected it can cause significant morbidity and mortality to the patient. Pregnancy in such a patient presents furthermore challenges and worsening of symptoms. Anaesthetic management of patients with TOF requires thorough understanding of anatomical defects and its physiological adaptations, and also the events and drugs that can alter the magnitude of R-L shunt. Problems in such patients are of chronic hypoxia, polycythemia and coagulopathy, CHF, embolism, episodic and reactive pulmonary vasoconstriction and altered acid base status. We present a case of uncorrected tetralogy of fallot TOF who underwent emergency caesarean section. Its hallmark anterior and superior infundibular septal displacement gives rise to the tetrad of ventricular septal defect, aortic override, infundibular obstruction, and right ventricular RV hypertrophy 2. Any disease complicated by severe maternal hypoxemia is likely to lead to miscarriage, poor fetal growth, preterm delivery or fetal death. There is a relationship between chronic hypoxemia and the polycythemia it causes with the outcome of pregnancy. With satisfactory surgical correction prior to pregnancy, maternal risks are decreased dramatically, and fetal environment is improved. Intracardiac repair has permitted survival into the childbearing years and excellent quality of life 6. Long-term complications usually relate to functional competence of the RV outflow tract and its secondary effects on ventricular and atrial myocardial function. Pulmonary regurgitation or stenosis may result in RV dysfunction and failure, progressive tricuspid valve regurgitation, atrial and ventricular arrhythmias, and sudden cardiac death 7. Case Report A 28 year female primigravida was posted for emergency cesarean section, having complaints of dyspnoea, early fatigability and history of cyanotic spells during heavy exertion before her pregnancy. She had history of taking beta blocker propranolol and digitalis off on. Her cesarean section was decided because of failed progression of labor because of cephalo-pelvic disproportion. Clubbing and murmur was present. As patient came in emergency her other investigations were not available at time of surgery. General anaesthesia was planned for her surgery. Ranitidine and Metaclopromide was given half an hour before surgery. Amoxicillin was also given for prophylaxis. Uterus contracted slowly spontaneously. On completion of surgery patient was reversed with neostigmine 0. She was further referred to cardiothoracic department. Discussion Maternal heart disease complicates 0. The complex of anatomic malformation results from an anterior displacement of the conoseptum toward the right ventricle creating a malalignment VSD and a narrowing of the outflow tract of the right ventricle RV 2. The aorta is displaced anteriorly, straddling the muscular septum and arising from both ventricles. The obstruction to outflow of the RV usually involves the infundibulum of the RV but can arise from the pulmonary valve, its annulus, the main pulmonary artery or even in the peripheral pulmonary arteries. Elevated pressures in the RV from outflow obstruction and exposure to systemic pressure from overriding aorta lead to compensatory RV hypertrophy. The main characteristic of TOF is cyanosis. Cyanosis can result from three separate Mechanisms. Inadequate pulmonary blood flow, right to left shunting or intrinsic pulmonary disease. In TOF, cyanosis results from a right-to-left shunt at the level of ventricles and inadequate pulmonary blood flow. Because of the outflow obstruction, blood ejected from RV crosses the VSD and enters the overriding aorta. This reduces the amount of pulmonary blood flow available for oxygenation and adds desaturated blood to the systemic circulation. Pressures in the right ventricle are near to the systemic pressure. The likelihood of a favorable outcome for the mother with TOF depends upon the functional cardiac capacity of the patient before pregnancy, other complications that further increase cardiac load, and quality of medical care provided throughout pregnancy

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and surgical correction of the anomaly before conception. Pregnant mothers with TOF are affected differently depending upon if they remain uncorrected, have palliative or definitive procedure or they have residual defects after these procedures. The principle danger for a pregnant woman with TOF is cardiac decompensation because of inability to meet the additional demands imposed by the physiological changes of pregnancy and parturition. If present, infection, hemorrhage and thrombo-embolism compound the risk. The cardiovascular changes of pregnancy may unmask residual or recurrent TOF in patients with corrective procedures, who have been asymptomatic throughout their life after TOF repair. Before successful intracardiac repair of TOF was introduced in the 1950s, few patients reached childbearing age, and successful pregnancy was uncommon. Pregnancies were characterized by spontaneous abortions, stillbirths, and premature deliveries. Chronic hypoxemia in such patients leads to adaptations to provide adequate tissue oxygenation i.e. Such adaptive mechanisms may limit cardiac reserve and O<sub>2</sub> delivery during stress. As no specific technique is suggested for such patients and anaesthesia carries considerable risk, the management should be based on avoiding changes that would increase the magnitude of R → L shunt, dehydration should be avoided to improve the circulating volume by decreasing the blood viscosity. Both general and regional techniques have been employed successfully in parturient with TOF. Regarding cesarean section, general anaesthesia GA is probably the technique of choice. GA with endotracheal intubation provides airway protection, eliminates work of breathing and may reduce oxygen consumption. The complications of controlled mechanical ventilation include decreased venous return as well as ventricular dysfunction, compression of pulmonary vessels, hypoxemia, hypo or hypercarbia and acidemia. The choice of anesthetic drugs may not be of prime importance. In patients with dynamic right ventricular outflow obstruction, increases in heart rate and contractility should be avoided as they will worsen the obstruction and cyanosis. Anesthetic drugs and adjuncts having vagolytic or sympathomimetic effects should preferably be avoided. Epidural catheter techniques offer continuous, titrated anaesthesia or analgesia. In this case we used general anaesthesia as patient came as emergency with known TOF but without taking any medicine regularly and with cardiac decompensation. She had not gone any palliative or corrective procedure. To achieve this ketamine was used for induction and phenylephrine to increase SVR. She was preloaded with 10 ml RL before induction. Patient was hyperventilated to maintain slightly alkalosis. Oxytocin was not used to avoid sudden increase in preload. Invasive monitoring CVP and arterial blood pressure could not be monitored in that case because of unavailability. Patient was managed properly. Patients with tetralogy of Fallot with pregnancy need special care by a team consisting of obstetrician, cardiologist and anesthesiologist among others.

Incidence of congenital heart disease: Surgical anatomy of tetralogy of Fallot. *J Thorac Cardiovasc Surg* ; Paediatric cardiac anaesthesia, 3rd ed. Sawhney H et al. Pregnancy and Congenital Heart Disease → maternal and fetal outcome. Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. *N Engl J Med* ; Pregnancy after surgical correction of tetralogy of Fallot. Bitsch M, Johansen C, et al. *Acta Obst Gyn Scand* ; Why Mothers Die The Confidential Enquiries into maternal deaths in the UK. Pregnancy in cyanotic congenital heart disease: Anaesthetic management of known case of tetralogy of fallot undergoing brain abscess drainage → A case report. *Indian J Anaesth* ; Congenital heart disease, Anaesthesia and coexisting disease, Churchill livingstone, 3rd ed. Anaesthesia for the obstetric patient with cardiac disease. *Clin Obst Gyn* ; Connolly, Martha Grogan, Naser M.

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## Chapter 5 : JOURNAL OF EVOLUTION OF MEDICAL AND DENTAL SCIENCES

*Tetralogy of fallot accounts for about 10% of CHD and about 50% succumb during first year of life. Children surviving this period present with hypoxia, cyanosis, polycythemia, coagulopathies, congestive heart failure and cyanotic spells.*

Find articles by Chitra Juwarkar Sidhesh S. Bharne Find articles by Sidhesh S. This article has been cited by other articles in PMC. Abstract Tetralogy of Fallot is the most common cyanotic congenital heart disease. Patient was a known case of heart disease since childhood who was advised surgery, but she had not undergone the same. She was not taking any medications at home. In the past, she had had three abortions and had undergone curettage twice. The anesthesia details were not known, but they were uneventful. On examination, she had cyanosis [ Figure 1 ], peripheral as well as central, and grade 2 clubbing [ Figure 2 ]. There was no variation of pulse and blood pressure in the extremities. Pedal edema was present. Cardiovascular system revealed a pansystolic murmur of grade 3 in the pulmonary as well as aortic areas, radiating all over the precordium. Rest of the examination was unremarkable. Her hemoglobin was Echocardiography showed a large subaortic ventricular septal defect with predominant right to left shunt. Right ventricular cavity was predominant with right ventricular hypertrophy. Biventricular function was within normal limits. There was moderate to severe pulmonary stenosis which was infundibular and valvular type. The pulmonary gradient was 68 mm. ECG showed sinus tachycardia with P pulmonale with poor R wave progression. Arterial blood gas analysis further confirmed shunting pH 7. Chest X-ray showed boot-shaped heart [ Figure 3 ].

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## Chapter 6 : Tetralogy of Fallot - Wikipedia

*Tetralogy of Fallot is the most common cyanotic congenital heart disease. We report the anesthetic management of a patient with uncorrected Fallot's tetralogy for Cesarean section.*

Tetralogy of Fallot Tetralogy of Fallot Tetralogy of Fallot is a combination of four congenital abnormalities. The four defects include a ventricular septal defect VSD , pulmonary valve stenosis, a misplaced aorta and a thickened right ventricular wall right ventricular hypertrophy. They usually result in an insufficient amount of oxygenated blood reaching the body. These defects, which affect the structure of the heart, cause oxygen-poor blood to flow out of the heart and to the rest of the body. Tetralogy of Fallot is often diagnosed during infancy or soon after. However, tetralogy of Fallot might not be detected until later in life in some adults, depending on the severity of the defects and symptoms. Tetralogy of Fallot care at Mayo Clinic Symptoms Tetralogy of Fallot symptoms vary, depending on the extent of obstruction of blood flow out of the right ventricle and into the lungs. Signs and symptoms may include: A bluish coloration of the skin caused by blood low in oxygen cyanosis Shortness of breath and rapid breathing, especially during feeding or exercise Loss of consciousness fainting Clubbing of fingers and toes " an abnormal, rounded shape of the nail bed Poor weight gain Tiring easily during play or exercise Irritability A heart murmur Tet spells Sometimes, babies who have tetralogy of Fallot will suddenly develop deep blue skin, nails and lips after crying or feeding, or when agitated. These episodes are called tet spells and are caused by a rapid drop in the amount of oxygen in the blood. Tet spells are most common in young infants, around 2 to 4 months old. Squatting increases blood flow to the lungs. When to see a doctor Seek medical help if you notice that your baby has the following symptoms: This helps increase blood flow to the lungs. Call or your local emergency number immediately. While factors such as poor maternal nutrition, viral illness or genetic disorders might increase the risk of this condition, in most cases the cause of tetralogy of Fallot is unknown. The four abnormalities that make up the tetralogy of Fallot include: Pulmonary valve stenosis is a narrowing of the pulmonary valve " the valve that separates the lower right chamber of the heart right ventricle from the main blood vessel leading to the lungs pulmonary artery. Narrowing constriction of the pulmonary valve reduces blood flow to the lungs. The narrowing might also affect the muscle beneath the pulmonary valve. A ventricular septal defect is a hole defect in the wall septum that separates the two lower chambers of the heart " the left and right ventricles. The hole allows deoxygenated blood in the right ventricle " blood that has circulated through the body and is returning to the lungs to replenish its oxygen supply " to flow into the left ventricle and mix with oxygenated blood fresh from the lungs. Blood from the left ventricle also flows back to the right ventricle in an inefficient manner. This ability for blood to flow through the ventricular septal defect reduces the supply of oxygenated blood to the body and eventually can weaken the heart. Normally the aorta " the main artery leading out to the body " branches off the left ventricle. In tetralogy of Fallot, the aorta is shifted slightly to the right and lies directly above the ventricular septal defect. In this position the aorta receives blood from both the right and left ventricles, mixing the oxygen-poor blood from the right ventricle with the oxygen-rich blood from the left ventricle. Over time this might cause the heart to stiffen, become weak and eventually fail. Risk factors While the exact cause of tetralogy of Fallot is unknown, various factors might increase the risk of a baby being born with this condition. These risk factors include: A viral illness during pregnancy, such as rubella German measles Alcoholism during pregnancy A mother older than age 40 A parent who has tetralogy of Fallot The presence of Down syndrome or DiGeorge syndrome Complications All babies who have tetralogy of Fallot need corrective surgery. Without treatment, your baby might not grow and develop properly. Your baby may also be at an increased risk of serious complications, such as infective endocarditis " an infection of the inner lining of the heart or heart valve caused by a bacterial infection. Untreated cases of tetralogy of Fallot usually develop severe complications over time, which might result in death or disability by early adulthood.

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## Chapter 7 : Tetralogy of Fallot - Symptoms and causes - Mayo Clinic

*Tetralogy of Fallot (TOF) is one of the most common congenital heart disorders (CHDs). This condition is classified as a cyanotic heart disorder, because tetralogy of Fallot results in an inadequate flow of blood to the lungs for oxygenation (right-to-left shunt) (see the following image).*

Advanced Search Congenital heart disease CHD is one of the most common inborn defects, occurring in approximately 0. The majority of these adults will require lifelong cardiological surveillance. GUCHD patients can broadly be divided into three categories: There are several reasons why patients may present with uncorrected lesions: Patients with balanced pulmonary and systemic circulations, as commonly occurs in patients with complex lesions, may remain relatively asymptomatic until the balance between pulmonary and systemic circulations is disturbed. Some patients will have previously been considered to be inoperable, and some patients may have come from overseas where facilities are unavailable. Paediatric cardiac surgery has changed enormously over the last 50 years. As a consequence, the pathophysiology of the patients is changing. Some palliative procedures have been abandoned, and increasingly there is a move towards the use of complex, sometimes staged, procedures for the correction of complex lesions. As they become adults, this is no longer appropriate. Increasing numbers of women are reaching reproductive age and requiring anaesthetic management during labour and delivery. Management of these women is based on knowledge of the physiological changes that occur during pregnancy, assessment of the existing degree of cardiovascular impairment and a detailed knowledge of the underlying pathophysiology. Some lesions are extremely well tolerated throughout pregnancy; others can decompensate disastrously. The effect of damage to the central nervous system and the success of ventricular preservation at the time of their previous procedures require careful consideration. In many patients in whom there has been a good outcome from cardiac surgery and in whom there is no evidence of late deterioration, conventional anaesthetic management is appropriate. In patients with functional limitation, the anaesthetic technique is modified to take account of the main problems currently presented by the patient. Sedative premedication to reduce oxygen consumption is popular, although care must be taken in patients with cyanotic heart disease and PVD. The benefits of regional anaesthesia, alone or in combination with general anaesthesia, need to be weighed on an individual basis. The benefits from good postoperative analgesia, and avoidance of activation of the sympathetic nervous system with consequent catecholamine release, need to be weighed against the effects of a reduction in systemic vascular resistance SVR. The circulatory effects of the i. All of the commonly used i. The need for invasive monitoring depends upon the nature of the surgery as well as the underlying cardiac lesion. Previous Blalockâ€™Tausig Bâ€™T shunt placement requires arterial pressure monitoring on the contralateral side. Central venous canulae pose a significant thromboembolic risk in patients with a Fontan type of circulation, and yet the information they provide may be extremely valuable. Pulmonary artery catheterization can be difficult because of anatomical abnormalities, and is not without risk in the patient with reactive PVD. Postoperative care of these patients is in either a high dependency unit or intensive care unit ICU. This approach may precipitate catastrophic cardiac failure in GUCHD patients with poor ventricular function. During IPPV, the duration of inspiration has a greater effect than the peak inspiratory pressure. Consequently, decreasing the duration of inspiration, and consequently increasing peak inspiratory pressure, is usually the best strategy to maximize pulmonary blood flow. Probably the greatest difference in management of GUCHD patients with complete mixing compared with normal patients is in the interpretation of a high saturation. Whilst this is a normal goal for the care of patients in the ICU, in patients with central mixing there is a hyperbolic relationship between arterial saturation and the ratio of pulmonary and systemic flows. Both the American Heart Association 20 and the European Society of Cardiology 41 clearly recommend that, for patients at high and moderate risk, antibiotic prophylaxis should be used when performing procedures associated with a risk of bacteraemia. Previously, labour and delivery have been regarded as high risk. In patients with limited pulmonary perfusion, avoidance

of perioperative dehydration, maintenance of SVR, control of PVR, and minimizing increases in oxygen consumption are central to a successful outcome. Regardless of the cause of hypoxaemia, it has profound haematological effects that affect many other organ systems. Polycythaemia is a compensatory response that improves oxygen transport at the expense of an increase in viscosity. Under most circumstances, the increase in viscosity is well tolerated, although undoubtedly these patients are at an increased risk of thrombosis and stroke. Multiple coagulation factor deficiencies are also common in patients with cyanotic CHD, producing a patient who is at risk of both spontaneous and excessive perioperative bleeding. The effect of chronic hypoxia on the heart is to induce myocardial dysfunction that usually manifests as a reduction in ventricular diastolic compliance and a reduction in myocardial reserve. The limitation of cardiac output may not be apparent at rest, but frequently there is a marked limitation in exercise tolerance. Chronic renal hypoxia produces marked glomerular abnormalities<sup>27</sup> that are associated with a reduction in glomerular filtration rate and result in a rise in plasma creatinine and urate. The latter is aggravated by the increase in red cell turnover. The hyperuricaemia is well tolerated and does not require intervention. These lesions result in pulmonary hypertension and right ventricular RV hypertrophy early in life. If this situation is allowed to persist, the changes in the pulmonary vascular tree become irreversible. At this stage, correction of the underlying lesion is not accompanied by a return of PAP to normal. The right ventricle is not designed to function against a high afterload, and ultimately will fail. Although many of the lesions likely to produce pulmonary hypertension are now corrected early in childhood, there still exists a sizable cohort of patients who underwent surgery in an era when definitive surgery was performed late. The aim of the management of these patients is to avoid factors that predispose to pulmonary hypertension and to reverse any reversible factors Table 4. Where practical, regional anaesthetic techniques are preferred. If general anaesthesia is required, controlled ventilation is almost mandatory. The Eisenmenger complex is characterised by a PVR greater than  $5 \text{ dynes cm}^{-2}$ , with a reversed or bidirectional shunt flow. The structural changes to the pulmonary vascular bed progress relentlessly, starting in childhood. Later there is medial hypertrophy of the more proximal muscular pulmonary arteries, associated with a rise in mean PAP. Finally there is a reduction in the number of distal pulmonary vessels accompanied by an increase in PVR. In fact many of them do surprisingly well: The actuarial survival is substantially better than that of patients with primary pulmonary hypertension. Ventricular function is usually well maintained, 40 dysfunction occurring most commonly in the setting of complex congenital heart disease. The development of symptomatic RV failure carries a poor prognosis. Fluid shifts and hypovolaemia, as can occur with haemorrhage, can result in a marked fall in cardiac output in a heart that is highly preload dependent. As a consequence of these physiological limitations, surgical procedures are associated with a high morbidity and mortality. This has been fuelled by the knowledge that maintenance of the SVR is vital. In the past, general anaesthesia has frequently been preferred. In addition to the type of anaesthetic, there are other equally important considerations. The risk of systemic embolization is not insignificant; all i. There are no outcome studies to guide the use of invasive cardiac monitoring. When a Bâ€™T shunt is inadequate or impossible, then a central shunt between the aorta and pulmonary artery may be the only method of providing tolerable pulmonary perfusion. It is difficult to regulate the blood flow through a central shunt, frequently it is excessive, and may give rise to pulmonary hypertension. The flow through a palliative shunt is largely determined by the size and length of the shunt, which are fixed, and the systemic driving pressure and PVR, which are under control of the anaesthetist. Measures that lower systemic arterial pressure in patients with a palliative arterial shunt can have disastrous effects on pulmonary perfusion and should be used with extreme caution. Arrhythmias Arrhythmias are the main reason for hospitalization of adults with GUCHD, 89 and account for over half of all emergency admissions. Risk factors for development of IART are older age at surgery and longer follow up. Changes in surgical technique, such as the development of the total cavopulmonary connection TCPC instead of the classical Fontan have been associated in a reduction in its incidence. Major risk factors for the development of VT are duration of follow up<sup>69</sup> and QRS prolongation to greater than ms, especially if associated with pulmonary regurgitation and RV

dilatation. Sudden late cardiac death remains a problem in GUCHD patients, occurring at a much greater incidence than in an aged matched population. Specific conditions It is impossible in the space available for this review to comprehensively cover the congenital lesions that are likely to be met in clinical practice. Detailed guidelines for the management of patients with CHD have been published following the Canadian Consensus Conference in 18 and by the European Society of Cardiology. In many cases, particularly those with complex lesions, referral to a regional centre is the best option. Atrial septal defects ASDs are subclassified according to their exact location; however, their physiological consequences essentially are independent of their site, depending on their size and the extent of any shunt. The behaviour of the shunt depends on the compliance of the two ventricles and the size of the defect. A 20 mm defect on the other hand is associated with a large shunt that can have considerable haemodynamic effects. The majority of ASDs are detected in childhood, although a significant minority are only diagnosed in adult life. PBF is increased and pulmonary hypertension develops with increasing age. A consequence of these physiological changes is dilatation of both left and right atria, the RV and the pulmonary arteries in order to accommodate the increased blood volume. Ultimately either the RV fails, or RV compliance falls, resulting in a reduction in the magnitude of shunting, or even flow reversal. Although pulmonary hypertension is quite common with increasing age, it is rare for the PVR to exceed  $5 \text{ dyn s cm}^{-2}$ . A few ASDs present in childhood with breathlessness, or even heart failure, but most are detected by echocardiography after a child has been referred for other reasons. In adult life, presentation commonly occurs because of breathlessness, atrial arrhythmias or heart failure, although some individuals remain essentially asymptomatic until shunt reversal occurs. Unfortunately later closure remains a risk for premature late death. Although small ASDs are generally considered benign, like a patent foramen ovale PFO they can allow paradoxical embolization to occur. There is, however, no doubt that in rare individuals, spontaneous cerebrovascular events related to a PFO can occur. Concern has been raised that venous air embolization during neurosurgical procedures may lead to paradoxical embolization in patients with a PFO. The era of transcatheter closure has now arrived, and today most ASDs can be closed percutaneously. Additionally, an ultimate conversion to a univentricular circulation, as opposed to transplantation, may be chosen for children with hypoplastic left heart syndrome. A univentricular circulation is inherently inefficient because of the recirculation of the pulmonary and systemic venous return. Since the late s, these patients have undergone palliative procedures in which the systemic venous blood is diverted to the pulmonary arteries, the single ventricle functioning as a systemic ventricle. Two classes of shunt have been developed. The first was the Glenn shunt in which the SVC was connected to the right pulmonary artery. This only supplied blood to the right lung, and has largely been superseded by the bidirectional Glenn shunt in which the SVC is anastomosed to the main pulmonary artery.