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RESEARCH ARTICLE

APPROACH TO CYANOTIC CONGENITAL HEART DISEASE IN CHILDREN: A SYSTEMATIC REVIEW

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Abstract

Objective: A growing number of research on the approach to children with cyanotic congenital heart diseasehave been undertaken; nevertheless, there is no clear consensus on which approach is superior to the other. The goal of this systematic review was to systematically review the available evidence on the approach to children with cyanotic congenital heart disease.

Methods: Authors began with recognizing the important examination proof that spots light on the approach to children with cyanotic congenital heart disease. We led electronic writing look in the accompanying data sets: Ovid Medline (2015-present), Ovid Medline Daily Update, Ovid Medline in process and other non-filed references, Ovid Embase (2015-present), The Cochrane Library (latest issue) and Web of Science. Just examinations in English language were incorporated. The precise selection was acted in close collaboration with a clinical examination curator.

Results: We included 8studies (956 participants). Although all certainty of evidence was good, approach to children with cyanotic congenital heart disease was similar among all studies. Use of cardiac echography was the modality of diagnosis among all studies. All studies emphasized on the importance of early detection for early intervention.

Conclusion: Early detection and treatment of cyanotic congenital heart disease (CCHD) is critical for positive outcomes. CCHD is a general term for a variety of diseases with different etiologies, which affect the clinical manifestations of CCHD. Totally abnormal total pulmonary venous connection (TAPVC) and transposition of the great arteries (TGA) are common in neonates. TOF and related disorders are characterized by squatting, cyanotic episodes, and a calm, resting chest without evidence of congestive heart failure, whereas transposition physiology is characterized by congestive heart failure with cyanosis.

Introduction:-

Congenital heart and great vascular abnormalities are the most prevalent severe congenital malformations [1-3], and they have a significant death rate in the first year of life [4]. Several studies with distinct demographics have found a frequency of 2 to 10 per 1,000 live births.

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Congenital heart disease (CHD) is defined as a gross anatomical abnormalities of the heart or intrathoracic great vessels that is either functionally significant or has the potential to do so [1]. CHD is regarded as the most serious congenital abnormalities and the main cause of death in the first year of life in the developed world [2,3]. However, data from poor nations is scarce. The prevalence of CHD at birth is estimated to be eight per 1000 live births [4,5]. Because of the large birth rate, the burden of CHD in emerging nations is anticipated to be considerable. Every year, it is predicted that about 180,000 infants in underdeveloped nations are born with CHD [6].

Recent advancements in cardiovascular diagnoses and treatment have boosted the survival of newborns and children with CHDs. Unfortunately, appropriate care for children with CHD is mostly limited to industrialized countries. It is critical to obtain solid information regarding the prevalence of CHD birth in the developing countries in order to plan and give better care to these patients. An previous research [7] from India reported on the prevalence of CHD at birth, however the diagnosis is mostly dependent on clinical evaluation.The usual clinical evaluation of infants has a low sensitivity for detecting CHD [8,9]. The gold standard for the diagnosis of CHD in infants is echocardiography with Doppler, which has a very high sensitivity and specificity [10,11,12].

The incidence and prevalence of cyanotic congenital heart disorders (CCHD) varies greatly throughout the world. It is responsible for one-third of all congenital cardiac disease [1, 4-5].

It affects offspring, everything being equal. Albeit the issue starts in utero, the time of show differs relying upon the sort of heart injury, how much blockage in the heart as well as pulmonary vasculature, and any attendant cardiovascular anomalies. TGA, for instance, may show from the get-go in the neonatal period, however TOF may not appear for the rest of the initial segment of the year. In more unfortunate countries with chronic weakness looking for conduct and restricted admittance to superb medical care, conclusion might be deferred until puberty in specific cases.

Regardless of well established interest in this disease, the particular reason for CCHD stays hazy [8-12].

Multidisciplinary care is furnished to young with CCHD [13-17]. Clinical treatment and careful fix of the irregularities might be consolidated. The monetary weight is significant, and it is shared by the family, the clinic, and the public authority. Getting medication and careful treatment is for the most part more costly than expected [18-21].The current review aimed to systematically review the available evidence on the approach to children with cyanotic congenital heart disease.

Methods:-

Review Question

This review seeks to systematically review the available evidence on the approach to children with cyanotic congenital heart disease. The specific review questions to be addressed are:
(1) What are the different approaches to children with cyanotic congenital heart disease?
(2) What are the differences between these approaches for children with cyanotic congenital heart disease?
(3) What is the prognosis among children with different types of cyanotic congenital heart disease?

Searches

We began with recognizing the important examination proof that spots light on the approaches available evidence about the management children with cyanotic congenital heart disease. We led electronic writing look in the accompanying data sets: Ovid Medline (2015 -present), Ovid Medline Daily Update, Ovid Medline in process and other non-filed references, Ovid Embase (2015 -present), The Cochrane Library (latest issue) and Web of Science. Just examinations in English language was incorporated. The precise selection was acted in close collaboration with a clinical examination curator.

Also, the bibliographies of any qualified articles recognized was checked for extra references and reference look were done for all included references utilizing ISI Web of Science.

We considered “published” articles to be compositions that showed up in peer-reviewedjournals. Articles present in grey literaturewere excluded from our review.

Types of studies to be included
We included articles covering how to coordinate different review plans in orderly review of seeks the latest updates on the approach to children with cyanotic congenital heart disease. We did exclude articles only providing prevalence and clinical characteristics of children with cyanotic congenital heart disease without focusing on the approach to these children.

We concentrated on the latest updates on the approach to cyanotic congenital heart disease. We included articles depicting sample sizes and articles that planned to sum up their outcomes to the population which test was drawn from. Case series and case reports were excluded from our search. Studies from all area all over the world were incorporated.

Participants
The systematic review included examinations with tests of children less than 12 years of age diagnosed with cyanotic congenital heart disease.

Searching key words
For every data set, looking through was led by utilizing a mix of the accompanying keywords: (cyanotic congenital heart disease OR guideline approach ORcyanosis ORheart murmur ORpulmonary atresia OR tricuspid atresia OR tetralogy of fallot OR critical pulmonary atresia OR hypoplastic left heart syndrome OR transposition of great arteries OR total anomalous pulmonary venous return OR systematic review).

We included examinations enrolling members in everyone as well as clinical settings. Studies were incorporated assuming they revealed the approach to children with cyanotic congenital heart disease. No comparator or control test size is required in the review to be incorporated.

Studies selection process
All list items were brought into an EndNote record. Two analysts evaluated titles and abstracts for their likely pertinence.

One reviewer freely screened titles and abstracts from the search and any articles that report approach to children with cyanotic congenital heart disease. We gained the full text of articles that possibly meet the eligibility criteria.

There was no geographical limit on the included studies. Just published articles in the English language will be incorporated.

Outcomes
Primary outcome
To systematically review the available evidence on the approach to children with cyanotic congenital heart disease.

Secondary outcome
To evaluate the clinical outcome and prognosis of children with cyanotic congenital heart disease.

Information extraction, (choice and coding)
Information was extracted from the included articles utilizing an electronic information extraction structure on Microsoft Access programming. Two reviewers freely extracted information, utilizing a standard information extraction structure which was created by the survey creators with the end goal of the review. The extraction structure incorporated the accompanying data:

1- Publication subtleties: title, authors, journal name, year and place of study, of distribution, country in which the review was led, sort of distribution, and wellspring of financing.
2- Study subtleties: concentrate on plan (cross-sectional, cohort, case-control), settings (clinical or population based), concentrate on transience (planned or review), patients' enlistment techniques (successive or non-continuous), the geographical area, year of information assortment and reaction rate, qualification (consideration and avoidance rules), name of appraisal tool(s), approval of evaluation tool(s).
3- Study members' subtleties: number of people reviewed/examined, population qualities including mean age (SD), and gender distribution, relationship status, demographic data.

Data management

A descriptive statistics is employed and relevant data are extracted from eligible studies and presented in tables. We then presented a narrative synthesis of the summary of the approach to children with cyanotic congenital heart disease.

Results:-

A total of 40 studies were identified in the search, all of them were assessed for eligibility, and 8articles were included in this review (Figure 1).

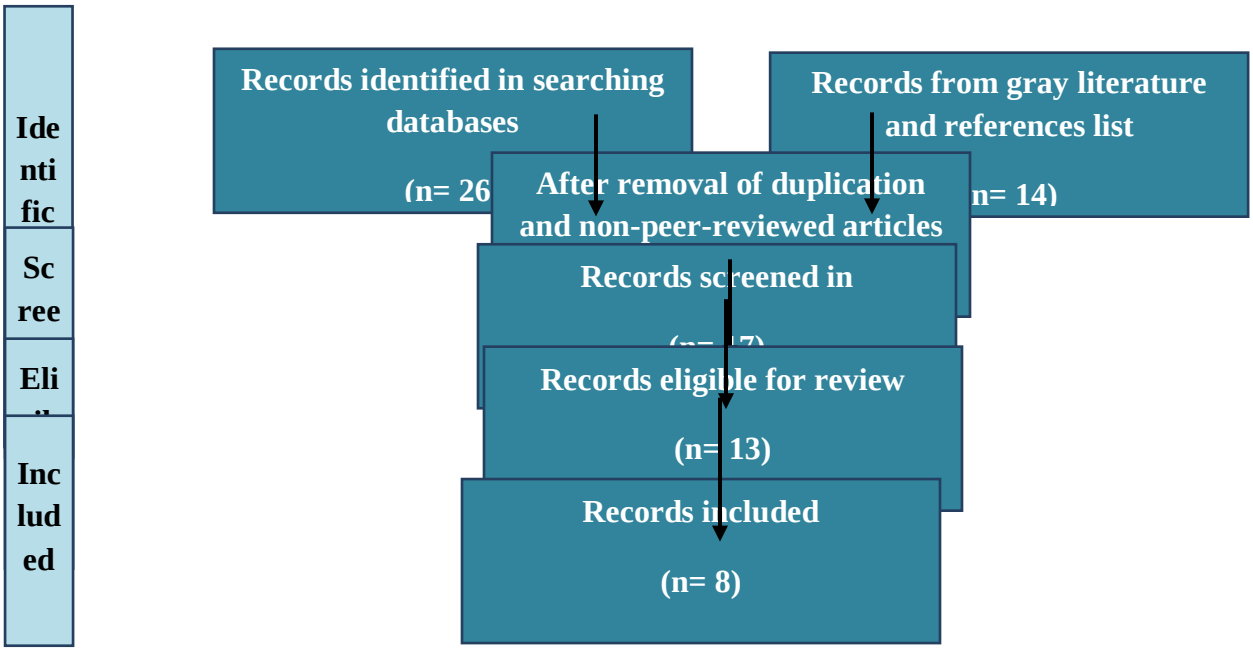

Figure 1:- Flow chart of selection process.

Congenital heart disease (CHD) affects 8-9 of 1000 births, and about 25% are classified as CHD. A second pregnancy after giving birth to a child with CHD, or if one parent is affected, increases the risk of CHD by 2% to 6%. The most common CCHD is tetralogy of Fallot (TOF) (5% of all CCHDs). Transposition of the great arteries (TGA) is the second most common CCHD (about 2% of all CCHDs) and the most common CCHD that appears within the first week of life. Cardiovascular disorders due to congenital malformations are estimated to cause 35% of neonatal deaths [22]. After prenatal echocardiography diagnosis of CCHD, specialized delivery room planning at a tertiary care facility is essential for timely assessment and treatment [23]. Table 1 summarizes the characteristics of the studies included.

Table 1:- Characteristics of RCTs studies included in this review.

Study	Year	Participants	Most common CCHD	Survival Analysis	
				% (95% CI)	Studies (I^2 %)
Wise-Faberowski [22]	2019	47	ToF (N= 31)	90.0 (89.0–91.0)	133 (84.1)
Bigdelian [23]	2018	125	VSD (N= 83)	88.0 (87.0–90.0)	70 (86.9)
Van De Bruaene [24]	2018	50	ToF (N= 24)	93.0 (91.0–94.0	63 (78.1)
Tulloh [25]	2014	87	ASD (N= 61)	81.0 (78.0–84.0)	110 (95.6)
Di Filippo [26]	2017	204	ToF (N= 107)	83.0 (79.0–86.0)	62 (95.6)
Bedair [27]	2019	304	TGA (N= 130)	79.0 (74.0–84.0)	48 (95.9)
Ismail [28]	2018	107	ToF (N= 67)	22.0 (17.0–26.0)	65 (96.9)
Nasir-Ahmad [29]	2018	32	ToF (N= 17)	18.0 (12.0–25.0)	30 (97.6)

If CCHD is suspected at birth and at the time of cardiac assessment, or if a pediatric cardiologist cannot respond immediately, appropriate stabilization, oxygen therapy, prostaglandin E1 infusion, and immediate evacuation to a

tertiary care hospital are required. Prostaglandin E1 is beneficial as a bridging treatment for subsequent surgery or cardiac surgery for tube-dependent lesions. About 25% of babies born with CHD may need heart surgery or other procedures to survive. In the TGA situation, balloon atrial septal defect causes proper blood mixing and, in the case of pulmonary hypertension, the pressure on the right side is relieved. Patients may undergo early orthodontic surgery or, rarely, be shunted before orthodontic surgery. After heart surgery, it is important to identify and treat the underlying causes of postoperative conditions such as: twenty four]. Vaccinations should be given on a regular basis. Patients who are candidates for cardiopulmonary bypass, heart, or heart-lung transplantation should carefully consider the timing of live viral immunity. During the respiratory syncytial virus season, infants with unresolved congenital heart disease and severe hemodynamic abnormalities should be protected from respiratory syncytial virus. According to a 2007 statement from the American Heart Association, prevention of subacute bacterial endocarditis (SAB) is recommended for patients undergoing dental surgery at high risk of adverse outcome (AHA). Other therapeutic concerns include managing iron deficiency anemia, frequent monitoring of excess erythrocytosis, and avoiding dehydration to reduce the risk of stroke. Parents of children with congenital heart disease should be counseled about possible heart problems in future children [25-26]. The one-year survival rate for newborns with CCHD has improved over time, but the mortality rate remains high. About 75% of newborns born with CCHD live to the age of one. About 69% of newborns with severe CHD are expected to live to the age of 18. Children with CHD are more likely to have developmental delay or disability, cardiac arrhythmias, heart failure, sudden cardiac arrest, or stroke [27-29].

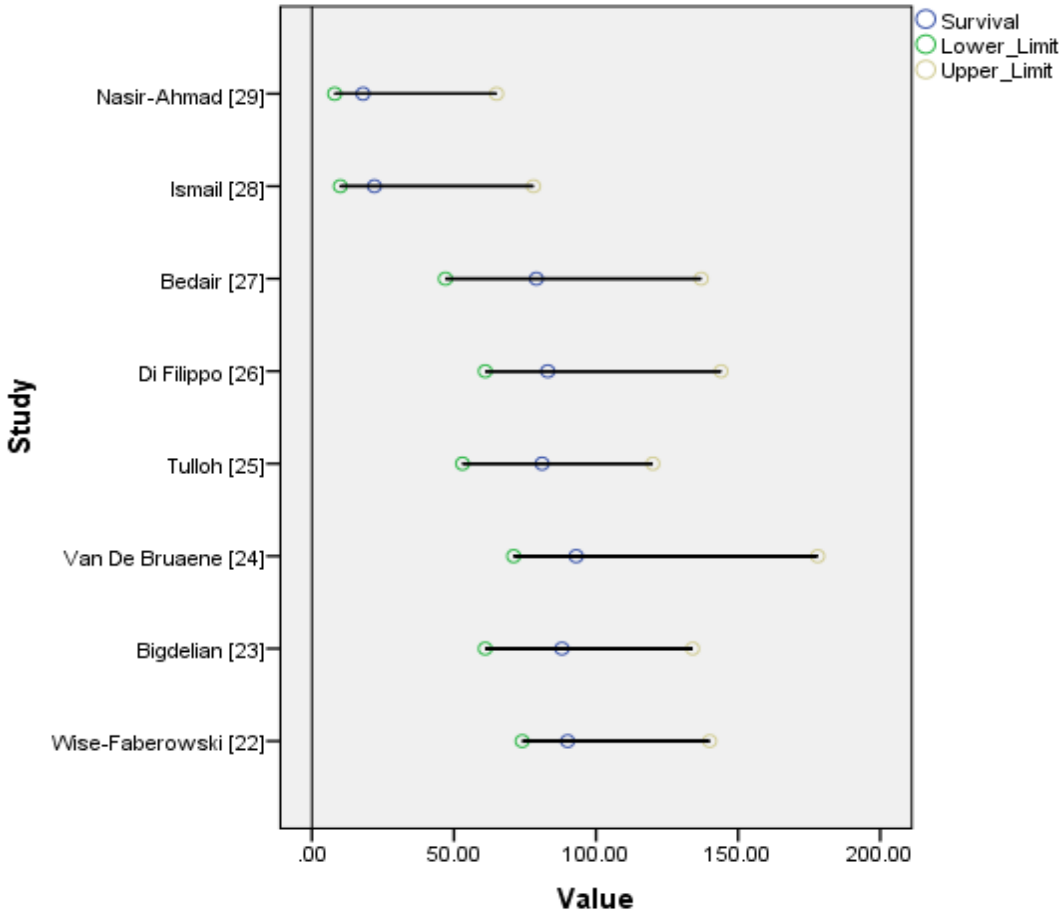

Figure 1:- Survival Analysis.

Discussion:-

The age at which a child is diagnosed with CCHD depends on the type of heart lesion and the severity of the lesion, other comorbidities, and other factors such as residence (Western countries or underdeveloped regions of the world). children with TGA, TAPVC, TA, persistent truncus, and DORV were diagnosed earlier than patients with TOF. However, the general age at diagnosis reported here was consistent with previous findings [17, 19, 21, 27]. The difference lies in the study design. Current studies focus only on CCHD, but previous reports cover all congenital

heart disorders. There were no reports based solely on CCHD. Related results show that most children are diagnosed with CCHD, and most CCHD diagnoses are made within 12 months and 1 month.

A possible explanation for the increase in the number of diagnoses in the first year could be the increased availability and experience of the facility in diagnosing heart disease. In Western countries, the diagnosis of CHD is made early in pregnancy or during the neonatal period, as opposed to reports from sub-Saharan Africa. In contrast, most cases in developing and emerging countries are not found until the end of childhood. Some cases are not diagnosed until the child is in elementary school or early adolescence. Possible explanations for delays in presentation and diagnosis include difficulty in assessing care by professionals, poverty, and poor health [28-30]. In the current study, the male-female ratio of CCHD was about the same. Several studies [20,21] have documented similar reports. Other researchers [17, 18, 25, 27] have observed an even distribution between men and women. A possible explanation for the conflicting findings is that the methodology and design of the study differed between the studies. There were differences in subject age, study duration, criteria used to select people, and so on. The current study focused on CCHD and was conducted over 9 years and 6 months, while other studies included non-cyanotic and cyanotic heart lesions, most of which were conducted over 5 years.

Regarding the distribution of various forms of cyanotic heart lesions in our study, TGA was the most common anomaly, followed by TOF. This is very different from other studies, given that TOF is widely recognized as the most common CCHD [31-33]. As a result, the distribution of certain congenital heart diseases is consistent throughout the region, regardless of geographic location or genetic composition. DORV and TOF were the second and third most common CCHDs. Previous studies have shown that TOF is followed by TGA [31-33], which is inconsistent with our findings. However, this may be due to a lack of information from other newspapers. This is because the number of isolated DORVs was slightly less than the number of TGAs. Other studies have identified far fewer cases of DORV than current studies. The massive rationale for current research is unknown. Possibility includes the longer duration of the current study and the diagnostic skills of the physician who evaluated the case. Again, these are speculative. Compared to previous results, the current study reports a significant number of cases of persistent truncus, TA and TAPVR. A possible reason is that the current study may have been conducted for a longer period of time than the previous study. Other less common heart diseases are single ventricle syndrome and hypoplastic left heart syndrome [34-38]. Cyanosis was the most common form of expression and justification for the evaluation of the heart. This is to be expected, considering that all patients had CCHD. However, some children became cyanotic, and medical history and physical examination suggested ACHD. Various clinical symptoms have been observed in certain studies. As mentioned in [39], some people have had symptoms such as brain abscess, stroke, and fatigue. It is well known that children with CHD exhibit a wide range of clinical symptoms and may require high suspicion indicators to make a diagnosis. Current research also provides information on the various subtypes of CCHD. TOF was the most common CCHD and accounted for just under half of all CCHDs [40]. It was predominantly male, with more than half of the children being cyanotic in the presentation. [41] The most common age group at diagnosis was 1 to 5 years. [42] Previous studies have shown that males predominate in TOF [23, 25, 29, 34, 35]. Kennedy etc. [36] reported equivalent age at the time of presentation in Malawi. This is later than the diagnosis during pregnancy in developed countries and much earlier in infancy. DORV, like TOF, showed male dominance, with cyanosis being the most common symptom [43]. However, unlike TOF, the majority of children were diagnosed when they were under one year old. Like DORV cases, TGA and TA cases are predominantly male, cyanosis is the most common symptom, and the majority of cases were diagnosed within 1 year [44].

TGA differs from the other two cases because it has the lowest median age at diagnosis. This is not surprising given that TGA is the most common cyanotic congenital heart disease in the neonatal period and the majority of cases are diagnosed in utero and in the neonatal period [2,31]. There was a slight difference between persistent truncus and other major CCHDs [45]. Cases of persistent truncus occurred mainly in women, but in TA, men and girls were evenly distributed. However, as with DORV, the majority of children with persistent truncus and TA were identified before the age of 1 year, with cyanosis being the most common symptom [5]. Children with CCHD receive interdisciplinary care [13]. You can combine medical treatment with surgical repair of abnormalities. The financial burden is considerable and is shared among families, hospitals and governments. Procurement of drugs and surgical treatments is generally more expensive than expected [14-16]. In most cases, surgical adjustment is required.

Conclusion:-

Early detection and treatment of cyanotic congenital heart disease (CCHD) is critical for positive outcomes. CCHD is a general term for a variety of diseases with different etiologies, which affect the clinical manifestations of

CCHD. Total anomalous pulmonary venous connection (TAPVC) and transposition of the great arteries (TGA) are common in neonates. TOF and related disorders are characterized by squatting, cyanotic episodes, and a calm, resting chest without evidence of congestive heart failure, whereas transposition physiology is characterized by congestive heart failure with cyanosis.The most common CCHD is tetralogy of Fallot (TOF) (5% of all CCHDs). Transposition of the great arteries (TGA) is the second most common CCHD (about 2% of all CCHDs) and the most common CCHD that appears within the first week of life.

Use of echocardiography is the best tool for diagnosing CHD. About 75% of newborns born with CCHD live to the age of one. About 69% of newborns with severe CHD are expected to live to the age of 18.

Cyanotic congenital heart illness usually manifests as during the neonatal period or early childhood. A thorough examination of indications and symptoms is critical in developing the management strategy. Syndromic traits can be used as diagnostic clues, thus a full body examination is required. The ECG and chest radiographs enhance the clinical symptoms and aid in the diagnosis. Once the kid is stabilized, or if intense cardiac care is required for stabilization, he or she should be sent to a pediatric cardiology center. If performed by a pediatric cardiologist, an echocardiogram will confirm the final diagnosis, and the child will be referred for surgical or catheter-based treatments as needed.

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