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Pediatric Arrhythmia Management in Congenital Heart Disease (CHD): A 20-Year Epidemiologic Study and Therapeutic Insights at Imam-Reza Hospital (2001-2020)

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Abstract

Background: Arrhythmia in children with congenital heart disease (CHD) can be due to abnormal anatomy, genetic mutations, hemodynamic abnormalities, primary myocardial disease, hypoxic tissue damage, or postoperative complications.

Objectives: This study aimed to investigate the epidemiological findings of children with various arrhythmias and their treatment outcomes.

Methods: The study was cross-sectional. The data were derived from 762 patients with arrhythmias, including tachyarrhythmias, bradyarrhythmias, and premature beats. The patient's demographic and clinical information, as well as their electrocardiogram (ECG), Holter monitor, echocardiography, and electrophysiology results, were collected and analyzed using SPSS software.

Results: The study found that 44.6% of the patients were female and 55.4% were male. The mean age of patients with tachyarrhythmias was 5.9 years, while those with bradyarrhythmias were 10 years old, and those with premature beats were 6.5 years old. The most common symptoms were palpitations, murmur, and slow pulse. The most common arrhythmias were supraventricular tachycardia (SVT) and premature atrial contractions (PACs).

Conclusion: The study revealed that cardiac abnormalities, including septal defects and valve abnormalities, were prevalent in patients with arrhythmias. Cardiomyopathy was associated with a higher risk of mortality. The study's findings emphasize the importance of paying attention to the presence of accompanying anomalies, palpitations, abnormal heart sounds, or bradycardia during the examination, as these are associated with an increased risk of mortality.

Keywords: Cardiac Arrhythmia, Congenital Heart Disease, Electrocardiography, Tachycardia, Bradycardia

1. Background

The prevalence of cardiac arrhythmias in children affects approximately 30,000 children annually. These arrhythmias can be due to congenital heart disease or occur in children with structurally normal hearts (1). The incidence of arrhythmias in pediatric emergency departments is approximately 55.1 per 100,000 visits, with sinus tachycardia being the most common, followed by supraventricular tachycardia (SVT), which accounts for around 13% of cases. Bradycardia comprises approximately 6% of all instances (2). Other prevalent types include atrioventricular reentrant tachycardia (AVRT), atrioventricular nodal reentrant tachycardia (AVNRT), ventricular tachycardia (VT), idiopathic VT, and congenital heart block. Neonatal arrhythmia is not uncommon and can affect infants with both normal hearts and CHD. Some arrhythmias are harmless and do not affect a child's life or health. However, some cases can lead to severe ventricular arrhythmia, with a heart rate exceeding 200 beats per minute in some patients. Without intervention, arrhythmias can cause permanent myocardial damage, leading to heart failure or even sudden death (3).

Arrhythmias often have sudden attacks in children, making them difficult to detect and diagnose. Electrocardiograms are often used for diagnosis, which can be inconvenient and costly. Pediatric cardiologists often prescribe Holter monitors, event monitors, or implantable loop recorders (ILRs) for evaluation. However, these devices provide a limited use period and sometimes require repeated use to record transient arrhythmias (4).

Tachycardia arrhythmia in some patients does not show obvious symptoms and can only be detected through physical examination. Some patients have shown signs of heart palpitations, chest pain, and even fainting in severe cases. Without proper treatment, tachyarrhythmia can alter hemodynamics, potentially cause myocardial ischemia and reduced cardiac function, and ultimately lead to cardiomyopathy (5).

Heart disease at birth not only affects anatomical defects but also causes electrical changes that lead to various arrhythmias. In addition, surgical corrections may also be associated with arrhythmias in CHD patients. Previous studies have reported that patients' arrhythmic substrates predispose them to arrhythmias associated with CHD, and manifestations and treatments differ between patients with normal hearts and CHD patients (6,7).

Cardiac channelopathy indicates arrhythmic conditions associated with genetic defects in ion channel genes. These include congenital long QT syndrome (LQTS), short QT syndrome (SQTS), catecholaminergic polymorphic VT (CPVT), and Brugada syndrome. Congenital LQTS is an inherited ion channel disorder associated with ventricular repolarization, characterized by syncope and sudden cardiac death, identifiable by polymorphic VT or torsade de pointes. SQTS patients present with sudden death, syncope, atrial tachycardia, and AF, occurring in patients aged 3 months to 70 years (8,9).

In the past, the focus of treating children's arrhythmias involved selecting appropriate antiarrhythmic medications based on the type of arrhythmia. However, this conventional therapeutic method can only alleviate symptoms but not provide a definitive cure. Additionally, the long-term use of antiarrhythmic drugs can seriously affect the patient's growth and development, limiting their widespread use (10).

Today, with the continuous advancement of interventional medicine, radiofrequency ablation has become an alternative to drug therapy for treating various arrhythmias in children (11).

Ablation with radiofrequency (RFA) has even become the preferred treatment method for many types of arrhythmias. Radiofrequency ablation utilizes radiofrequency energy to produce and direct

thermal energy, thereby destroying critical pathways in the heart that can cause arrhythmias and preventing their occurrence (12). Thus, this procedure will inevitably have unavoidable adverse effects on the myocardium and may even cause damage to it (13).

2. Objectives

This epidemiological study examines the prevalence of various arrhythmias in children and the treatment outcomes at Imam Reza Hospital in Mashhad, Iran, over 20 years. The study aims to gain insights into the epidemiology and management of pediatric arrhythmias, providing valuable data for healthcare providers to enhance patient care and inform effective treatment strategies.

3. Methods

The current study was conducted at the pediatric cardiology department of Imam Reza Hospital in Mashhad, which currently serves as the region's sole referral center for pediatric cardiac care. This study aims to collect and analyze data on the prevalence of various arrhythmias and their treatment outcomes in a selected 20-year period. In addition to arrhythmia types and treatment results, relevant demographic information and cardiac diagnostic findings from procedures such as echocardiography, 24-hour Holter monitoring, electrocardiography, cardiac catheterization, and electrophysiology studies were gathered as extensively as possible.

The inclusion criteria encompass all children referred and treated for arrhythmia on an outpatient or inpatient basis. The exclusion criteria included incomplete records, age younger than 1 month or older than 18 years, and lack of follow-up by a pediatric cardiologist.

The statistical analyses were conducted using IBM SPSS Statistics, version 22. The data were compared between the groups via Pearson's chi-square or Fisher's exact tests. A P value of less than 0.5 was considered statistically significant.

4. Findings

In this study, 857 patients who met the inclusion criteria were initially included. However, 95 patients (11%) were excluded based on the exclusion criteria. The reasons for exclusion included: incomplete records (4.7%, 41 patients), age outside the range of 1 month to 18 years (2.9%, 25 patients), and lack of follow-up by a pediatric cardiologist (1.7%, 15 patients).

After accounting for the excluded cases, 762 patients met the inclusion criteria and were categorized into three groups:

Tachyarrhythmias (38.1%, 291 patients), Bradyarrhythmias (9.9%, 76 patients), Premature atrial and ventricular contractions (51.8%, 395 patients).

4.1. Tachyarrhythmias:

In this category, 291 patients were included. The electrocardiographic findings of these patients can be elaborated as follows:

Ventricular tachycardia (4.1%, 12 patients), Atrial flutter (1.4%, four patients), Atrial fibrillation (1%, three patients), Wolff-Parkinson-White (2.1%, six patients), AV reentrant tachycardia (2.1%, six patients), Atrioventricular nodal reentrant tachycardia (2.4%, seven patients), Junctional tachycardia (2.4%, seven patients), Non-specified (84.5%, 246 patients).

In the analysis of treatment outcomes for the 291 patients in the tachyarrhythmia group, 187 patients received medical therapy, with 5 cases (2.7%) experiencing recurrence and two patients (1%) developing complications (1 case with possible cardiac arrest and 1 case with circulatory complications). Overall, 180 patients (96.3%) continue to be managed with medical therapy.

No reports of mortality, cardiac arrest, or

complications were recorded for the 29 patients who underwent ablation.

Fisher's exact test demonstrated statistically significant associations between mortality and the following factors: cyanosis, hematologic disorders, genetic disorders, and restrictive or dilated cardiomyopathy ($P < 0.05$). No significant relationship was found between mortality and other demographic factors, laboratory findings, echocardiography results, or arrhythmia types ($P > 0.05$).

4.2. Bradyarrhythmia:

A total of 76 patients were part of this study group. Among these patients with bradyarrhythmias, 53 patients (72.6%) exhibited heart block, including five patients (6.85%) with second-degree heart block and 48 patients (63.1%) with third-degree heart block.

Additionally, 23 patients (30.2%) experienced sinus bradycardia.

Various treatment approaches were employed:

- 29 patients (38.1%) received medical therapy.
- 15 patients (19.7%) underwent interventional procedures.
- 18 patients (23.6%) underwent surgical treatment.
- Detailed information about the treatment outcomes was not available for 14 patients.

Out of the 76 bradyarrhythmia patients, 33 (43.3%) had pacemakers, including 26 patients (78.7%) had primary pacemakers, 21 patients (63.3%) had epicardial leads, 11 patients (33.3%) had endocardial leads, and one patient (3.4%) initially had an epicardial lead, later replaced with an endocardial lead.

Complications occurred in 8 patients (24.2%), including lead dysfunction in 3 patients (9.1%) and local pacemaker site infection in 3 patients (9.1%).

Death or cardiac arrest was observed in only four patients (5.3%):

- 2 patients (2.65%) experienced postoperative death.
- 1 patient suffered from severe local pacemaker site infection.
- 1 patient experienced a pacemaker malfunction.
- 1 patient had Heart failure following pulmonary hypertension.

In the bradyarrhythmia group, Fisher's exact test revealed a statistically significant association between mortality and the following factors: second-degree heart block, ventricular septal defects, and pulmonary hypertension ($P < 0.05$). No significant relationship was found between mortality and other demographic factors, laboratory findings, or echocardiography results ($P > 0.05$).

4.3. Premature atrial and ventricular contractions:

In the analysis, 395 patients with premature atrial and ventricular contractions were studied:

- 266 patients (67.3%) had premature atrial contractions.
- One hundred patients (25.3%) had premature ventricular contractions.
- Twenty-nine patients (7.3%) had both types of premature contractions.

Treatment outcomes in this patient group showed that 272 patients (68.9%) received medical therapy, while 123 patients (31.1%) did not experience recurrence and did not require long-term medication. Also, mortality was observed in only six patients (1.5%).

Using Fisher's exact test, a statistically significant association was found between mortality and the presence of dilated or restrictive cardiomyopathy ($P < 0.05$). However, no significant relationship was found between mortality and other demographic factors, laboratory findings, or echocardiography results ($P > 0.05$). Table 1 shows the abnormal echocardiographic findings in the studied patients.

Table 1: The abnormal echocardiographic findings based on types of arrhythmias in all patients.

Type of Arrhythmia	Premature atrial and ventricular contraction (395 Patients)	Brady-arrhythmia (76 Patients)	Tachy-arrhythmia (291 Patients)
Septal defects			
Ventricular septal defect	41 (10.4%)	17 (22.4%)	17 (5.8%)
Atrial septal defect	17 (4.3%)	15 (19.7%)	23 (7.9%)
Atrioventricular septal defect	4 (1%)	4 (5.3%)	1 (0.3%)
Arterial abnormality	0 (0%)	0 (0%)	0 (0%)
Patent ductus arteriosus	28 (7.1%)	27 (35.5%)	5 (5.2%)
Right-sided obstructive lesions			
Tetralogy of Fallot	14 (3.5%)	4 (5.3%)	6 (2.1%)
Pulmonary valve stenosis	23 (5.8%)	13 (17.1%)	9 (3.1%)
Pulmonary atresia	8 (2%)	0 (0%)	5 (1.7%)
Pulmonary vessel abnormality			
Pulmonary hypertension	9 (2.3%)	11 (14.5%)	6 (2.1%)
Left-sided obstructive lesions			
Aortic stenosis	5 (1.3%)	0 (0%)	2 (0.7%)
Coarctation of the aorta	3 (0.8%)	0 (0%)	2 (0.7%)
Myocardial abnormality			
Hypertrophic cardiomyopathy	0 (0%)	0 (0%)	3 (1%)
Restrictive cardiomyopathy	3 (0.8%)	1 (1.3%)	1 (0.3%)
Dilated cardiomyopathy	7 (1.8%)	4 (5.3%)	8 (2.7%)
Complex (Double Outlet Right Ventricle, Transposition of great arteries, Heterotaxia)	4 (1%)	4 (5.3%)	2 (0.7%)
Tricuspid valve abnormality			
Ebstein anomaly	19 (4.8%)	0 (0%)	16 (5.5%)
Mitral valve abnormality			
Congenital MR OR MS	34 (8.6%)	0 (0%)	21 (7.2%)
Minor Abnormality			
FMV/MVP Without MR or trace MR	166 (42%)	15 (19.7%)	92 (31.7%)
Patent foramen ovale	56 (14.2%)	11 (14.5%)	38 (13.1%)
Persistent left superior vena cava	1 (0.3%)	1 (1.3%)	1 (0.3%)

5. Discussion

This study examined the demographic, clinical, and echocardiographic characteristics of pediatric patients with various types of arrhythmias. The findings offer insights into the prevalence and characteristics of arrhythmias in this population, highlighting the factors associated with mortality.

The study divided patients into three categories: tachyarrhythmia, bradyarrhythmia, and premature atrial and ventricular contractions.

In the tachyarrhythmia group, male patients predominated, with an average age of 5.9 years. The majority had negative parental consanguinity, and the cardiac causes were the most common etiology.

The study's findings revealed that the most frequent minor echocardiographic abnormalities were patent ductus arteriosus (PDA) and patent foramen ovale (PFO).

On the other hand, supraventricular tachycardia (SVT) was the most common arrhythmia observed.

Fisher's exact test showed that cyanosis, hematologic disorders, genetic disorders, and restrictive or dilated cardiomyopathy had significant associations with mortality ($P < 0.05$).

In the bradyarrhythmia group, no gender dominance was observed. Also, the average age was 10 years, and most patients had negative parental consanguinity. In these patients, cardiac causes were the most common etiology.

The study's results revealed that the most frequent minor echocardiographic abnormalities were ventricular septal defects (VSDs) and floppy mitral valves (FMVs). On the other hand, the premature atrial contractions (PAC) were most commonly seen.

Fisher's exact test showed that second-

degree heart block, ventricular septal defects, and ventricular septal rupture had significant associations with mortality (P < 0.05).

In the premature atrial and ventricular contractions group, no gender dominance was observed. Most patients had negative parental consanguinity, and cardiac causes were the most common etiology. In this group, the most frequent minor echocardiographic abnormalities were atrial septal defects (ASDs) and floppy mitral valves (FMVs). It is worth mentioning that premature atrial contractions (PACs) were most commonly observed. Fisher's exact test showed that restrictive and dilated cardiomyopathy had significant associations with mortality (P < 0.05).

An examination of interventional procedures performed on patients throughout the 20-year study:

The study evaluated the effects of interventional treatments on patients with tachyarrhythmias (ablation) and bradyarrhythmias (pacemaker implantation). Patients were first categorized into 5-year intervals, based on the range of 2001 to 2020.

A significant increase in ablation therapy was observed during the final 5 years of the study, particularly in the last 5 years. Additionally, there was an increase in pacemaker implantations for bradyarrhythmias during the study period.

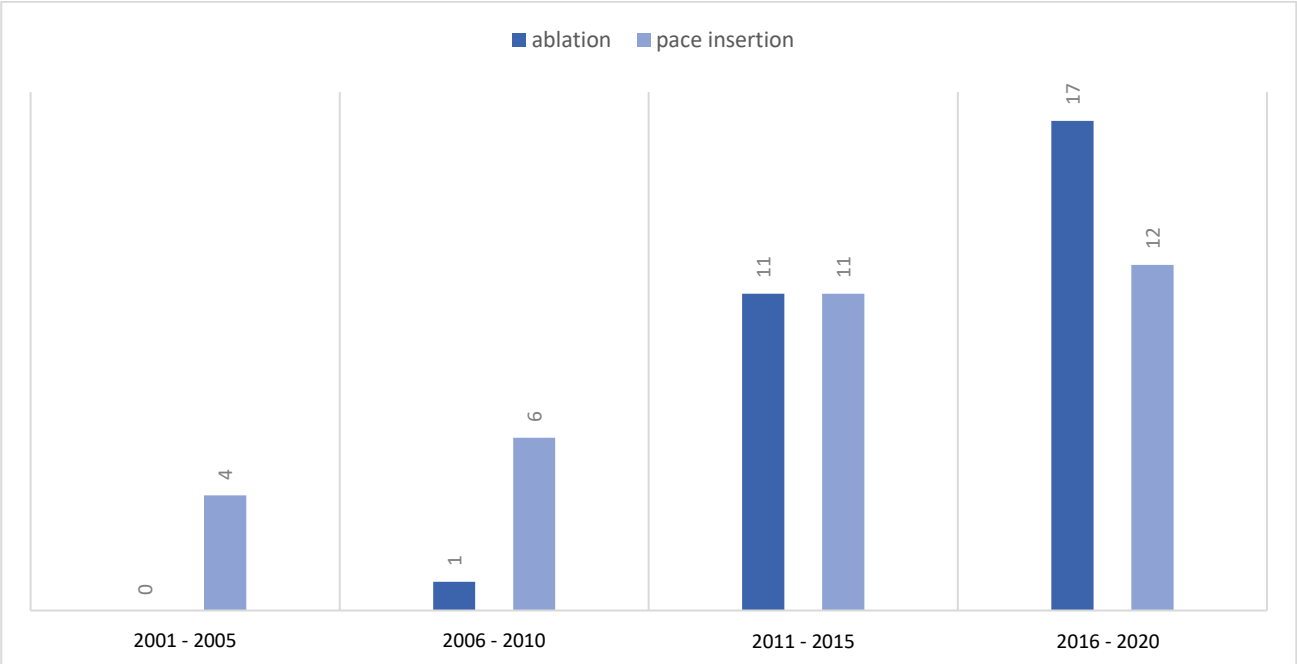

Figure 1: Frequency of Interventional procedures in Different Time Intervals throughout the Study years.

In a study by Talwar and colleagues on arrhythmias occurring in children after cardiac surgery, out of 224 pediatric patients who underwent heart surgery from September 2013 to July 2014, 24 patients were excluded because these procedures were performed without the use of cardiopulmonary bypass. Fifteen patients (7.5%) developed arrhythmias, with the most common arrhythmia being junctional ectopic tachycardia (JET) in 7 patients (46.6%), followed by supraventricular tachycardia (SVT) in 5 patients (33.3%). All cases of JET occurred within 24 hours of admission to the intensive care unit. Five of the seven patients with JET responded to conventional interventions, while two required amiodarone administration. They concluded arrhythmia, particularly JET, rarely occurs in children following cardiac surgery. Apart from a longer time for sinus rhythm recovery, no specific predictors were identified. It appears that the cause of post-cardiac surgery arrhythmias in children is multifactorial and warrants further

study with more significant patient populations (14).

For comparison, the most common arrhythmia observed in our study was SVT. This difference may be due to differences in the characteristics of the studied populations, highlighting the need for further research in our country.

In Isik et al.'s research, which was conducted as a series of case studies on neonatal arrhythmias, a retrospective study was performed on the birth process, and cases were identified during their stay in the neonatal intensive care unit (NICU) due to arrhythmia diagnoses in two NICUs in Turkey from May 2011 to June 2013. Seventeen neonates diagnosed with arrhythmia were identified. The incidence of NA in the two NICUs was 0.4% and 0.3%, and in the overall study population, it was 0.37%. The average gestational age was 37 weeks (6,14). In nine neonates (53%), fetal arrhythmia (FA) was diagnosed in the last week of pregnancy.
The distribution of NA types is as follows:

Six (35%) supraventricular tachycardia (SVT), Six (35%) premature atrial contractions (PACs), Two (11%) premature ventricular contractions (PVCs), Two (11%) multiple arrhythmias, such as PAC + SVT and PVC + AV block, One (5%) case of atrioventricular block (AV block). One patient had Wolff-Parkinson-White syndrome. NA was associated with congenital heart defects in five cases (15).

In the study by Badri and colleagues on the rhythms of hospitalized children in the NICU, a random sample of 457 infants was examined to identify the prevalence, types, and risk factors of cardiac arrhythmias. ECG lead 12 was monitored in all infants. One-quarter of the infants with normal ECGs (n = 100) underwent Holter studies, and all infants with abnormal ECGs (n = 39) also underwent Holter studies. Out of the 100 infants presumed to be free of arrhythmias based on ECG, nine infants showed arrhythmias during the Holter studies. When comparing Holter's findings with maternal, infant, and fetal risk factors, arrhythmias were significantly associated with male gender, higher maternal age, lower glucose levels, maternal smoking, abnormal umbilical cord insertions, and the use of β-2 adrenergic therapy. Dopamine administration was not associated with arrhythmias. The conclusion was that arrhythmias in the NICU are more common than in the general infant population. ECG monitoring had a sensitivity of only 89% compared to Holter monitoring (16).

In the study conducted by Rękawek et al., which examined the risk factors for postoperative arrhythmias in 402 patients aged 1 day to 18 years (mean 29.5 months) who underwent surgery, the detection and treatment of early postoperative arrhythmias were evaluated. Arrhythmias occurred in 57 out of 402 patients (14.2%). The most common types of arrhythmias were junctional ectopic tachycardia (21%), supraventricular tachycardia (15%), and ventricular premature beats (6%). In a stepwise logistic regression analysis, only a higher Aristotle score was statistically significant ($P < 0.001$), while body weight ($P = 0.62$) and age ($P = 0.0188$) were identified as significant in a single-variable analysis. In the stepwise multivariable logistic regression analysis, only the higher Aristotle basic complexity score was statistically significant ($P < 0.001$), while body weight ($P = 0.62$) and age ($P = 0.40$) were not (17). In our study, among the tachyarrhythmia subtypes, restrictive and dilated cardiomyopathies, as well as genetic and hematologic disorders, were significantly associated with mortality. In the group with bradyarrhythmias, second-degree heart block, ventricular defects, and pulmonary artery hypertension were also significantly associated with an increased risk of mortality.

In another study conducted by Hok and colleagues, all patients under 18 years of age diagnosed with congenital heart disease were examined. The study population consisted of 232 patients, with Ebstein's Anomaly (n=44), septal defects (n=39), and single-ventricle

heart (n=36) being the most common diagnoses. The most common arrhythmia mechanism was AVNRT in 109 cases, and the arrhythmia type was non-specific in 56 cases. They concluded that children with congenital heart diseases exhibit a wide range of arrhythmia mechanisms. They stated that despite the recurrence of new mechanisms after successful treatment, successful ablation treatment can be performed (18).

In another study conducted in Saudi Arabia from 2015 to 2020, 821 patients under the age of 18 were examined after undergoing heart surgery. Out of these, 140 patients experienced arrhythmia post-surgery, with the most common types being JET (51.4%), AV block (27.1%), and supraventricular tachycardia (10%). It was also found that most arrhythmia cases (79.3%) occurred within 24 hours after the heart surgery (19).

In another study, 580 children were examined after heart surgery until they were discharged from the hospital. Of these, 51 patients (8.8%) experienced arrhythmia at an average age of 2.3 ± 1.7 years. The most common arrhythmias were supraventricular tachycardia (41.1%), JET (23.5%), and complete AV block (19.6%). Ultimately, 15 patients (29.4%) experienced sustained arrhythmia, with at least 7 cases resulting in death due to treatment-resistant arrhythmia (20).

For comparison, the overall mortality rate was lower in the groups examined in our study, and the most common type of arrhythmia was SVT.

6. Conclusion

Congenital heart block and supraventricular tachycardias are among the most common arrhythmias observed in children. The prevalence of these cardiac rhythm disorders may be higher between the ages of 5.9 to 10 years. Parental consanguinity does not influence the occurrence of arrhythmias. Patients with concomitant anomalies, such as cardiac enlargement, heart murmurs, or bradycardia, require increased attention, especially when the primary referral reason is a cardiac defect. Cardiomyopathies can significantly increase the risk of sudden cardiac events and mortality in these patients. Abnormalities in the heart's conduction system and structure frequently accompany the emergence of cardiac arrhythmias.

In conclusion, this study underscores the complexity of pediatric arrhythmias and the need for a multidisciplinary approach to diagnosis, treatment, and long-term follow-up. Future research should focus on optimizing treatment protocols, reducing complications, and exploring genetic and molecular therapies to address the underlying causes of arrhythmias in children. By advancing our understanding and managing these conditions, we can improve the quality of life and outcomes for affected children and their families.

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