Published in final edited form as:

Am J Cardiol. 2016 September 15; 118(6): 906–911. doi:10.1016/j.amjcard.2016.06.034.

National Trends in Hospitalizations of Adults with Tetralogy of Fallot

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Abstract

The population of adults with tetralogy of Fallot (TOF) is growing, and it is not known how the changes in age distribution, treatment strategies and prevalence of comorbidities impact their interaction with the healthcare system. We sought to analyze the frequency and reasons for hospital admissions over the past decade. We extracted serial cross-sectional data from the United States Nationwide Inpatient Sample on hospitalizations including the diagnostic code for TOF from 2000 to 2011. From 2000–2011, there were 20,545 admissions for individuals with TOF, with a steady increase in annual number. The most common primary admission diagnoses were heart failure (HF; 17%), arrhythmias (atrial 10%, ventricular 6%), pneumonia (9%) and device complications (7%). The rates of comorbidities increased significantly, particularly diabetes (4.5% to 8.1%), obesity (2.1% to 6.5%), hypertension and renal disease. The number of pulmonic valve replacements increased (6.8% to 11.3% of TOF admissions, $p < 0.001$), with a rise in median age at surgery from 16 to 19 years old ($p = 0.036$). The cost per TOF admission was more than double that of non-congenital HF admissions and rose significantly, reaching $\$21,800 \pm \$46,000$ in 2011. In conclusion, hospitalized patients with TOF have become significantly more medically complex and are growing in number. The rise in the prevalence of obesity, hypertension and diabetes in this young population supports the need for prevention efforts focused on modifiable risk factors, in addition to HF and arrhythmia treatment. The increase in cost of care calls for further analysis of areas in which efficiency can be increased to ensure high quality of care and lifelong follow-up of patients with TOF.

Keywords

Tetralogy of Fallot; Cardiac Risk Factors; Supraventricular arrhythmia; Heart failure; Health care delivery

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect. While surgical correction in infancy and childhood pioneered in the last 60 years leads to a high rate of event-free survival, adolescents and adults with TOF are at increased risk for hospital admission due to late sequelae of those operations¹, and they need to be cared for by both general and specialized congenital physicians. Care of adults with TOF was dramatically changed in the last 2 decades with recognition of the deleterious effect of chronic pulmonary valve regurgitation, timing of pulmonary valve replacement (PVR) based on right ventricular size and function, clinical heart failure (HF) and risk of sudden cardiac death^{2–6}, and the introduction of percutaneous PVR. The effects on hospital admissions and resource use of an aging TOF population, as well as new evidence regarding the optimal timing and methods for PVR, have not been investigated. In this study, we utilize a comprehensive national sample of hospital admissions to assess trends in the rate of TOF admissions, reasons for hospitalization, cost of care and outcomes in the United States.

METHODS

We extracted data on TOF admissions from 2000 to 2011 from the Nationwide Inpatient Sample (NIS), part of the Healthcare Cost and Utilization Project⁷. The NIS is a database of diagnostic codes and de-identified data from a stratified sample approximating 20% of inpatient admissions in the United States. The Agency for Healthcare Research and Quality, which supports the database, provides sampling weights, which are then used to calculate national estimates⁷. Adolescents and adults (over the age of 10) admitted between 2000 and 2011 with an International Classification of Diseases-9th revision code for TOF (745.2) were included, similar to prior studies^{8–10}, and compared to the total number of admissions in the NIS. We analyzed demographic information available in the NIS, which includes age at admission, gender, year of admission, and Agency of Healthcare Research and Quality comorbidities which are based on Elixhauser's comorbidity measures, which include HF, chronic pulmonary disease, coagulopathy, diabetes, hypertension, liver disease, obesity, renal failure. PVR was assessed using the following surgical and percutaneous procedure codes: 33475, 35.07, 35.08, 35.25, 35.26; we excluded patients with a Rastelli procedure (35.83, 35.92). Electrophysiology procedures were selected according to the codes for pacemaker (37.80–37.89, 00.50–00.54, 33206–33208), lead revision (37.7), cardioversion (99.60–99.69, 92961), MAZE procedure (37.33–37.34), defibrillator (37.9–37.98, 33240–33249), electrophysiology testing (37.26–37.29, 93619–93624, 93285–93299) and ablation (37.34, 93650–93652). The primary diagnosis at admission was extracted for all patients; the secondary diagnosis was used for patients with TOF listed as primary. Admissions with the primary diagnosis code of HF (428) or arrhythmia (427) in TOF patients were compared with admissions with the same primary admission code but without a congenital diagnosis code, in the same time period. From the year 2001 onward, data on hospital charges per admission was extracted from the NIS, and converted to costs based on Healthcare Cost and Utilization Project Cost-to-Charges ratio files, which contain hospital- and year-specific data. Our cross-sectional study of observational data conforms with the Strengthening the Reporting of Observational Studies in Epidemiology guidelines. Summary statistics were reported as means with standard deviations (SD) for continuous variables and medians and

interquartile ranges (IQR) for non-normally distributed continuous data. P-values for the summary measures and estimates of the National counts were obtained using proc surveyfreq and surveyreg in SAS that accounts for the complex sampling of the NIS. Missing data was very rare for the measures of interest (<1%). The linear trend and the Mantel-Haenszel trend tests were used respectively for continuous and categorical variables to compare annual trends between time periods of 2000–2003, 2004–2007, and 2008–2011. Multiple linear regression was used to analyze the relationship between comorbidities and admission cost. Analyses were conducted using SAS software version 9.3 (SAS Institute Inc., Cary, North Carolina). A p-value < 0.05 was considered statistically significant.

RESULTS

There were 20,545 admissions for individuals with TOF overall between 2000–2011, indexed to the population of the United States (Table 1). The number of admissions increased over the study period at a similar pace as the overall admission rate in the NIS. The most common admission diagnoses were HF, arrhythmias and pulmonary disease, when excluding TOF itself and other anatomic diagnoses (such as “pulmonary valve disease”) (Table 2). The most common comorbidity (which are listed in a distinct field from the primary diagnosis at admission) was HF. There was, however, a significant increase in the rate of arrhythmias, hypertension, diabetes, obesity, renal and hepatic disease (Figure 1a), and the Charlson Comorbidity Index (from 0.7+/- 0.9 to 0.9+/- 1.0, p<0.001). The rates of obesity increased in adolescents and adults up to age 65; the population of young adults, aged 18–40, had the most marked increase in comorbidities (Figure 1b).

There were 1891 admissions for PVR, with a doubling of the annual rate (when compared to the total number of TOF admissions, trend p-value<0.001) during the interval studied (Figure 1a). There were no patients with billing codes for percutaneous PVR (35.07, 35.08) captured in this time period as these codes were implemented in October 2011; percutaneous PVR prior to then were done as part of clinical trials, and possibly used the surgical PVR codes. Patients admitted for PVR were less medically complex, as measured by the Charlson Comorbidity Index, than the general TOF population, but their complexity did increase over the study period (0.15 +/- 0.39 to 0.31 +/- 0.62, p=0.031). There was a significant shift towards PVR being performed in large hospitals (Table 1; as defined by the NIS, specific to region and academic status¹¹).

The average inflation-adjusted cost per admission increased from \$17,500 ±27,200 in 2000 to \$21,800 ±46,000 in 2011 (p=0.016; expressed in 2011 US dollars) in the population overall (Figure 2), double the cost of admissions for arrhythmia or HF in the non-congenital population in the NIS (which respectively were 10,200 ± 15 to 11,000± 16,000, and 9,800 ± 15,000 to 11,000 ± 17,000 in 2000 to 2011). The charges billed per admission were higher and had an increasing gap compared to the actually reimbursed costs described above (\$35,000 to nearly \$70,000 from 2000 to 2011). The median length of stay was stable at 3 days (IQR 2–6) for overall TOF hospitalizations and 5 days (IQR 5–7) for PVR admissions, with a trend towards decreased post-operative length of stay over the study period that did not reach statistical significance. A regression analysis showed that PVR was most strongly correlated factor with an increase in cost of the admission in the population overall; on

average, it added \$32,403 (95% CI \$27,826, \$36,979, $p<0.001$) to the total cost of the admission. In an exploratory multivariate analysis of adults between 18 and 40 years-old, an admission for arrhythmia, surgery during the admission and coagulopathy were correlated with an increase in cost, while a history of lung disease was inversely related to cost, and admission for HF, a history of diabetes or hypertension were not significantly independently associated with cost.

DISCUSSION

In this study, we demonstrate a significant rise in comorbidities, cardiovascular procedures, and cost of hospitalization in individuals admitted with TOF. The annual number of hospitalizations of patients with TOF is increasing and continues the trend seen in 1998–2005⁸. Improved survival, longer life expectancy¹², and an aging TOF population likely contribute to this increased number of hospitalizations. The overall rate of comorbidities, as reflected by the Charlson Comorbidity Index, has been increasing in both patients admitted for PVR and for the TOF group overall in this study, and parallels the increase in cost of hospitalization. Renal and hepatic disease are becoming more common, possibly related to a worse cardiovascular status and the increase in rates of diabetes and hypertension; this underscores the need for increased awareness and assessment of multiorgan disease. The rate of obesity tripled and that of diabetes and hypertension nearly doubled over the study period ($p<0.001$) in adults aged 18–40, even though the absolute numbers remain low. While our study population is only the subset of TOF patients in the United States who incurred a hospitalization in the last decade, this is the first report of rates in a large and diverse population. The focus on their congenital cardiovascular pathology at the detriment of other less serious conditions, and a tradition of exercise limitation, puts our patients at particularly high risk, especially in this era of a metabolic syndrome epidemic. With the implementation of ACHD-specific guidelines and movement to follow patients at ACHD centers of expertise^{6,13}, there also comes greater responsibility to consider the multiple comorbidities that will affect health. Exercise prescription, for which cardiopulmonary stress tests is a useful guide¹⁴, complements a team-based approach that includes primary care coordination, nutrition and activity assessment and counseling.

One in 4 hospitalized patients with TOF in this study had HF listed as a comorbidity. HF hospitalizations are known to be associated with high resource utilization and increased risk of readmission in the general cardiology literature, and with poor outcomes in adults with congenital heart disease¹⁵. In addition to a team-based approach to closely counsel and follow outpatients, earlier diagnosis with more sensitive and objective measures, such as biomarkers and cardiopulmonary exercise testing, is needed¹⁶. While the high reimbursement for HF admissions may be a motivator to include it as a diagnosis and could lead to over-reporting in administrative databases, the proportion of admissions due to HF in this analysis are similar to that reported in other studies, ranging from 20% at age 15 to 80% at age 50¹⁷.

This study demonstrates that the rate of PVR in patients with TOF, compared to all TOF admissions, has doubled over the past decade, particularly in adults. While the true incidence of PVR cannot be estimated from this data as the national prevalence of TOF is not known,

the rates are similar to previous data from an experienced single center in the United Kingdom¹⁸. This trend likely reflects practice-changing publications beginning in the 1990s on the consequences of pulmonary valve regurgitation and strategies to optimize timing of PVR, as well as the availability of percutaneous valves^{3,4,19}. The improvement in surgical technique and post-operative care, as well as national trends to operate on patients with more comorbidities²⁰, has likely now made surgery an option for older adults with TOF. The use of imaging markers of right ventricular dysfunction may have also identified and prompted a surgical referral in middle-aged adults before they progressed towards clinical HF and multi-organ dysfunction, which would have previously precluded surgery. Mortality during PVR admissions in this study was lower than in previous reports and similar to contemporary data^{18,21,22}, perhaps reflecting improvements in patient selection, surgical planning and approach (including better right ventricle protection during bypass) and postoperative care.

The cost per TOF admission is close to double that of contemporary non-congenital HF and arrhythmia admissions, comparators of interest since they are the most common causes for admission in patients with TOF and high resource use admissions within cardiology. While significantly associated with higher cost, admissions for PVR have had a stable cost and represent only 11% of all admissions. The observed shift towards admission and surgeries being performed in larger hospitals, which matches the national trend of concentration of specialized resources, is a likely significant contributor to the rise in hospital charges and cost; care in tertiary centers has however also been associated with improved outcomes in CHD patients²³. Non-elective admissions, government insurance, HF diagnosis, surgical complexity, renal failure and procedural complications have in previous studies been found to be strongly associated with higher resource use in adults with congenital heart disease¹⁰; common use of high-cost imaging tests is also likely to contribute and should be further evaluated. The sharp rise in hospital charges for the admission- i.e. the billing from the hospital-has outstripped the cost, which represents the actual reimbursement made to the hospital, at a rate that is not seen in contemporary patients with non-ACHD HF²⁴. Importantly, it signals a rising burden placed on patients with high deductibles or no medical insurance, which is increasingly at risk of being marginalized and lost to care²⁵.

The use of national level databases allows for a broader assessment of the effects of evolution in demographics of the population and changing practice patterns; it however carries several limitations. The total prevalence of TOF in the United States is not known, so the number of admissions recorded in the NIS cannot be put in the context of the size of the at-risk population. The NIS carries de-identified data and does not link multiple admissions for 1 patient; outcomes outside of the index admission are therefore not known and a cause-effect relationship between comorbidities or procedures and outcomes cannot be established. Full details of the procedures performed during the hospitalization is not available. In addition, administrative data can be of limited accuracy due to possible errors in billing and coding, and we do not have access to the primary records to verify the admission diagnoses. This database has however been recently used in several studies in ACHD^{8,26}, and we obtained congruent results to single center detailed studies on the incidence of surgery¹⁸ and rate of renal disease and diabetes²⁷⁻²⁹.

Acknowledgments

Funding Sources: Dr. Stefanescu Schmidt was supported in the manuscript preparation by the National Institutes of Health T32HL007604 Training Grant in Cardiovascular Research. The manuscript contents are solely the responsibility of the authors and do not necessarily represent the official views of National Institutes of Health.

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Figure 1.

Figure 1a: Trends in comorbidities

*trend $p < 0.0001$ ** trend $p = 0.0009$ for atrial arrhythmias and $p = 0.018$ for ventricular arrhythmias

Figure 1b: Rates of select comorbidities by age group

*trend $p < 0.0001$ *trend $p = 0.002$ *** trend $p = 0.026$ + trend $p = 0.018$ Other p -values > 0.1

Figure 2. Cost for TOF admissions, by reason for admission

HF: heart failure

Other p-values >0.1

Table 1

Demographics of TOF admissions

Characteristics	Overall	2000–2003	2004–2007	2008–2011	p-value
Number of admissions, indexed to U.S. population	20,545	6260	7113	7172	0.456 *
Mean age at admission (95% CI)	31.9 (15–48.8)	30.6 (15.2–46)	32.3 (15.8–48.8)	32.5 (16.3–48.7)	0.002
Proportion of adults (age 18)	75.7%	72.4%	76.1%	78.1%	<0.001
Female gender	52.1%	54.0%	52.1%	50.4%	0.062
Elective admission	33.7%	34.6%	33.4%	33.6%	0.737
Mortality during the admission	3.1%	3.2%	3.4%	2.8%	0.578
Location and teaching status of hospital					
Rural	6.8%	9.3%	5.9%	5.4%	0.036
Urban non-teaching	21%	20%	23%	20%	
Urban Teaching	72%	70%	71%	74%	
Disposition of patient					
Home	79.6%	80.9%	79.0%	79.1%	0.618
Home with services	7.0%	5.8%	7.0%	8.1%	
Transfer to short term hospital or rehab	9.6%	9.7%	9.8%	9.3%	
Discharge against medical advice	0.6%	0.5%	0.6%	0.6%	
Comorbidities					
Hypertension	13.4%	8.8%	12.2%	16.4%	<0.001
Diabetes mellitus	5.9%	4.5%	4.3%	8.1%	<0.001
Obesity ^{**}	4.0%	2.1%	2.2%	6.5%	<0.001
Congestive heart failure	24.3%	25.3%	23.3%	24.6%	0.734
Procedures during hospitalization					

Characteristics	Overall	2000–2003	2004–2007	2008–2011	p-value
PVR (% of total TOF admissions)	9.1%	6.8%	9.0%	11.3%	<0.001
Mean age at surgery (years, 95% CI)	23.5 (10.6–36.4)	20.7 (9.5–31.9)	24.9 (11–38.8)	23.7 (10.9–36.5)	0.075
Age>18 at PVR	52%	41%	54%	66%	0.037
Mortality during the PVR admission	0.8%	2.3%	0%	0.6%	NS
Proportion of PVR performed in urban academic hospital with >325 beds	66.8%	45.5%	60.8%	82.6%	<0.001
Electrophysiology procedure performed during admission	11.2%	10.4%	12.1%	11.0%	NS

* p-value for comparison with all admissions in the NIS, which are increasing as well.

** defined with diagnostic codes 286.0–286.9, 278–278.03, 649.1–649.14, 793.91, V85.3–85.39, V85.41–85.45, V85.54.

Abbreviations:

TOF: Tetralogy of Fallot

U.S.: United States

PVR: Pulmonary valve replacement

Table 2

Twenty most common primary admission diagnoses for TOF patients, grouped by categories

Diagnosis	% of TOF admissions
Valvular or structural disease *	16.6%
Heart failure	10.1%
Pulmonary disease	9.7%
Complication of surgery or device	6.8%
Supraventricular tachycardia	6.8%
Ventricular tachycardia	3.2%
Neurological disease	2.8%
Pregnancy and delivery	1.8%
Endocarditis	1.5%
Syncope	1.3%
Renal disease	1.1%
Chest pain	1.0%
History of cardiac surgery	0.8%
Gastrointestinal disease	0.6%
Hematologic disease	0.5%
Coronary artery disease	0.5%
DiGeorge syndrome	0.5%
Other **	34.4%

Percentages described are as proportion of from the patients admitted with the 20 most common diagnoses, excluding TOF.

* Includes pulmonary valve disease, pulmonary artery anomaly, pulmonary heart disease, atrial septal defect, ventricular septal defect.

** Includes diagnoses with individual prevalence of <0.5%

TOF: Tetralogy of Fallot