Objective: We sought to characterize the natural history of aortic root dilatation and aortic regurgitation (AR) in tetralogy of Fallot (TOF) and its variants.

Methods: This was a single-center, retrospective review of all TOF patients who underwent primary repair from 1/2000-12/2015, survived to discharge, and had ≥2 post-discharge echocardiograms. TOF anatomy was categorized as TOF-pulmonary stenosis (TOF-PS) or TOF-Variant (TOF-V, including TOF-pulmonary atresia [TOF-PA], TOF-PA-major aortopulmonary collateral arteries [TOF-PA-MAPCA], TOF-double outlet right ventricle, TOF-absent pulmonary valve syndrome [TOF-APVS], and TOF-complete atroventricular canal). Echocardiographically-determined aortic diameters and derived z-scores were measured at the annulus, sinus of Valsalva (SoV), and sinotubular junction (STJ) just before repair (baseline) and throughout clinical follow-up. Linear mixed effects models, comparing TOF-PS and TOF-V patients, assessed trends in aortic diameters over time.

Results: Of 624 patients meeting entry criteria, median age at repair was 3.0 (IQR 1.4-5.0) months and 386 (61.9%) had TOF-PS. Median postoperative follow-up was 13.9 (IQR 10.0-17.8; range 4.4-22.1) years. Each patient had an average of 15.6 +/- 9.4 post-discharge echocardiograms. There were 2 (0.3%) cases of aortic valve/root replacement and no cases of aortic dissection; 146 (23.4%) and 10 (1.6%) patients developed ?mild and ?moderate AR, respectively. The mean, linearized rate of aortic growth was: annulus, 0.09 +/- 0.05 cm/year; SoV, 0.13 +/- 0.06 cm/year; STJ, 0.11 +/- 0.06 cm/year (Fig A). Root z-scores remained largely unchanged over time (Fig B). TOF-V demonstrated larger annular (p=0.012), SoV (p<0.001), and STJ (p=0.018) diameters at baseline, as well as greater annular (p=0.039), SoV (p=0.001), and STJ (p=0.001) growth over time, relative to TOF-PS (Fig C). These differences were largely driven by TOF-PA-MAPCA and TOF-APVS. The rates of annular (p<0.001), SoV (p=0.003), and STJ (p=0.002) dilatation were higher (along with diameters at each 5-year increment) in patients that developed ?mild AR or required aortic valve/root surgery throughout the follow-up period (Fig D).

Conclusions: Aortic root z-scores in TOF remain stable through the second decade of life. Patients with TOF-V (especially TOF-PA-MAPCA and TOF-APVS) may be at relatively greater risk of pathologic root dilatation over time. Further longitudinal follow-up is warranted in this cohort.

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