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ORIGINAL ARTICLE

Aortic dilatation after tetralogy of Fallot repair: A ghost from the past or a problem in the future?

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KEYWORDS
Aortic dilatation;
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Abstract
Introduction and Aims: Intrinsic aortopathy can lead to dilatation late after tetralogy of Fallot (TOF) repair. Its extent and prevalence are not known. We aimed to assess aortic dimensions and elasticity and to find predictors of aortic dilatation.
Methods: A total of 126 adults were prospectively included after TOF repair and compared to 63 gender- and age-matched controls. Transthoracic echocardiography was used to assess aortic diameters at the level of the sinuses of Valsalva and ascending aorta and aortic dilatation was defined as z-score >+2. M-mode parameters of the ascending aorta were used to calculate strain, distensibility and stiffness index.
Results: TOF patients (mean age 30±9 years; 52% male) had a complete repair at a median age of five (2-49) years; mean follow-up time since repair was 23±7 years. The prevalence of aortic dilatation at the sinuses of Valsalva and ascending aorta was 29% and 24%, respectively. Compared to controls, TOF patients had a higher ascending aorta z-score, lower strain (6.4% [0.0-61.5] vs. 15.2% [0.0-45.0], p<0.01) and higher stiffness index (7.3 [0.8-23.6] vs. 3.1 [0.9-14.1], p<0.01). On multivariate analysis male gender was strongly associated with sinuses of Valsalva dilatation (odds ratio 6.3, 95% confidence interval 1.5-26.3, p=0.01).
Conclusions: The prevalence of aortic dilatation late after TOF repair is significant, with a larger and stiffer ascending aorta. Male gender appears to influence aortic root dilatation. This aortopathy requires careful follow-up in order to prevent future complications.

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PALAVRAS-CHAVE
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Tetralogia de Fallot;
Ecocardiografia transtorácica

Dilatação da aorta na tetralogia de Fallot operada: um fantasma do passado ou um problema no futuro?

Resumo
Introdução e objetivos: Uma possível aortopatia intrínseca poderá condicionar dilatação tardia da aorta na tetralogia de Fallot operada. A sua prevalência e extensão não estão definidas. Pretendemos avaliar as dimensões e a elasticidade da aorta e encontrar preditores da dilatação aórtica.
Métodos: Incluímos prospetivamente 126 adultos operados a tetralogia de Fallot e comparamos com 63 controlos. Avaliamos por ecocardiografia transtorácica os diâmetros dos seios de Valsalva e da aorta ascendente e definimos dilatação aórtica para z-score > +2. Parâmetros modo M da aorta ascendente foram usados para calcular *strain*, distensibilidade e índice de rigidez.
Resultados: Doentes com tetralogia de Fallot (idade média 30 ± 9 anos; 52% homens) foram operados com uma idade mediana de 5 (2-49) anos; tempo médio de seguimento desde a cirurgia 23 ± 7 anos. A prevalência de dilatação dos seios de Valsalva e da aorta ascendente foi 29% e 24%, respetivamente. Comparado aos controlos, os doentes com tetralogia de Fallot apresentaram maior z-score da aorta ascendente, menor *strain* (6,4 [0,0-61,5] *versus* 15,2 [0,0-45,0]%; p < 0,01) e maior índice de rigidez (7,3 [0,8-23,6] *versus* 3,1 [0,9-14,1]; p < 0,01). Na análise multivariada o sexo masculino associou-se significativamente à dilatação dos seios de Valsalva (*odds ratio* 6,3, intervalo de confiança de 95% 1,5-26,3; p = 0,01).
Conclusões: Na tetralogia de Fallot operada há uma prevalência significativa de dilatação tardia da aorta, com a aorta ascendente maior e mais rígida. O sexo masculino parece influenciar a dilatação da raiz da aorta. Esta aortopatia requer um seguimento cuidadoso para evitar complicações futuras.

List of abbreviations

2D	two-dimensional
AAo	ascending aorta
AAoZ	ascending aorta z-score
Ad	aortic diastolic diameter
Ao	aortic
AoZ	Ao root z-score
As	aortic systolic diameter
BP	blood pressure
BSA	body surface area
CI	confidence interval
D	distensibility
DBP	diastolic blood pressure
GUCH	grown-up congenital heart
LV	left ventricular
LVEF	left ventricular ejection fraction
OR	odds ratio
PP	pulse pressure
SAC	systemic arterial compliance
SBP	systolic blood pressure
SD	standard deviation
SoV	sinuses of Valsalva
STJ	sinotubular junction
SV	stroke volume
SVI	stroke volume index
TOF	tetralogy of Fallot
TTE	transthoracic echocardiography

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease in which survival into adulthood is common. The guidelines recommend regular long-term follow-up in specialized grown-up congenital heart (GUCH) centers.[1]

Major residual lesions after TOF repair occur at the level of the right ventricular outflow tract, including obstruction or aneurysm, and at the pulmonary valve due to regurgitation or stenosis. In addition, as first described by Capelli et al.,[2] there is an increasing awareness that aortic (Ao) dilatation can develop late after TOF repair. Interestingly, Ao dilatation is a fetal feature of TOF that tends to disappear after early surgical repair[3] without a palliative systemic-to-pulmonary shunt, thus preventing long-standing volume overload on the overriding aorta. Ao dilatation may be a disorder of the past, not reflecting the current surgical era.[4] Nevertheless, for many reasons, including anatomical and technical surgical issues, not all patients will benefit from early repair. Additionally, aortic histological abnormalities present since infancy in TOF, especially in the Ao root and ascending aorta (AAo) vascular wall, can contribute to Ao dilatation.[5,6] This intrinsic aortopathy can lead to Ao dilatation late after TOF repair, but its extent and prevalence are not known. We aimed to assess proximal thoracic aorta dimensions and elasticity by transthoracic echocardiography, in TOF patients and normal controls, and to find possible predictors of Ao dilatation.