patients sent for echocardiography during a specified period of time. Reasons for presentation were innocent heart murmurs, a family history of congenital heart disease, rule-out of cardiac side effects of non-cardiac medications, among others.

4. Patient populations. While comparing study populations, we would be interested to know the age of the patients and relatives studied by Fazio et al. Presumably most of them were adults. By whom criteria did patients enter the registry; was it hospital admissions only? How many NCVM patients were entered with and without congenital heart disease? NCVM subpopulations may carry a different cardio-vascular risk.

5. Incidence. We do not know the total number of patients entered into the highly specialized Italian registry. But certainly, the number of NCVM patients entered in 1 year (>230) is remarkable; as is the number of first-degree relatives detected with NCVM (48/31). It has been repeatedly stated that NCVM appears to have been previously under-diagnosed all together.

Pretty much all existing data on NCVM has been prone to a selection bias. We do not know the total number of patients entered into the highly specialized Italian registry. But certainly, the number of NCVM patients entered in 1 year (>230) is remarkable; as is the number of first-degree relatives detected with NCVM (48/31). It has been repeatedly stated that NCVM appears to have been previously under-diagnosed all together.1,2,5

Patient populations

References


Christian Lilje
Kinderkardiologie, Univ. Herzzentrum Martinistr. 52, D-20246 Hamburg Germany; and Ped. Cardiology, Tulane University 1430 Tulane Ave New Orleans, LA 70112 USA
Tel: +49 40 42803 3718 fax: +49 40 42803 6826 E-mail address: lilje@uke.uni-hamburg.de
Vit Rázek
Kinderkardiologie, Herzzentrum Univ. Leipzig Leipzig Germany
James J. Joyce
Ped. Cardiovasc. Center University of Florida Jacksonville USA
Thomas Rau
Pharmakologie/Toxikologie Univ.-Klinikum Hamburg-Eppendorf Hamburg Germany
Barbara F. Finckh
Klin. Pathologie/Pädiatrie Univ.-Klinikum Hamburg-Eppendorf Hamburg Germany
Florian Weiss
Diagnost. Radiologie Univ.-Klinikum Hamburg-Eppendorf Hamburg Germany
Christian R. Habermann
Diagnost. Radiologie Univ.-Klinikum Hamburg-Eppendorf Hamburg Germany
Janet C. Rice
Biostatistics Tulane University New Orleans, USA
Jochen Weil
Kinderkardiologie Univ. Herzzentrum Hamburg Germany

Septal alcohol ablation in hypertrophic obstructive cardiomyopathy: improving a myocardial scar

We read with great interest the article by van Dockum et al.1 on the improvement of systolic myocardial function of the left ventricular (LV) lateral (free) wall in patients with hypertrophic cardiomyopathy (HCM) after alcohol septal ablation (ASA). Using cardiac magnetic resonance (CMR) tissue tagging and three-dimensional strain analysis, the authors found that both maximum end-systolic strain index and systolic strain index rate improved significantly in remote myocardium.

This report shows for the first time that the reduction of the LV outflow tract gradient in symptomatic patients with obstructive HCM treated with ASA is associated with the improvement in intramural systolic function in the lateral wall remote from the ablated area. Although this is an interesting finding, there is a main point to be addressed in relation with the procedure. In Figure 1, there is a clear demonstration of a gross gadolinium late myocardial hyperenhancement in the interventricular septum attributable to the procedure, although there is no report of direct comparison with pre-procedural gadolinium myocardial enhancement in the same patient. It would be very interesting if myocardial hyperenhancement data derived by CMR before and after ASA could be provided by the authors. Such data would be very helpful to estimate the impact of ASA on the development of new fibrosis superimposed on an already existing one.

The most dramatic event in HCM is sudden death attributable to arrhythmogenic substrate owing to cardiac fibrosis. Cell death with subsequent healing and replacement fibrosis induced by ASA eventually leads to an increase in the already existing myocardial fibrosis, creating a substrate more prone to arrhythmic events. In other words, we are trying to improve patient’s symptoms by generating a scar tissue that may be deleterious long life, especially for young subjects. Data on sudden death after ASA is lacking. Therefore, as stated by Maron,2 avoidance of septal ablation in young patients is probably prudent, especially if the surgical option is feasible.

References

1. van Dockum WG, Kuijer JPA, Groote MJW, ten Cate FJ, ten Berg JM, Beek AM, Twisk JWR, Marcus JT, Visser CA, van Rossum AC. Septal ablation in hypertrophic obstructive cardiomyopathy improves systolic myocardial function in the lateral (free) wall: a follow-up study using CMR tissue tagging and 3D strain analysis. Eur Heart J 2006;27:2833–2839.


Georgios K. Efthimiadis
Cardiology Department AHEPA General Hospital Stilp. Kiriakidi 1, 54 637 Aristotle University of Thessaloniki.
We thank Efthimiadis et al. for their comments on our article concerning the improvement of systolic myocardial function in the lateral (free) wall in hypertrophic obstructive cardiomyopathy (HOCM) after alcohol septal ablation (ASA), which was studied using CMR tissue tagging and 3D strain analysis.1 In a previous work, we have demonstrated that contrast-enhanced imaging data derived in ~60% of the study-group pre-ASA after administration of gadolinium-DTPA contained only small pre-existing foci of delayed myocardial hyperenhancement, representing myocardial fibrosis and other pathological changes in the myocardial wall (e.g. disarray, inflammation, oedema, myo-lysis, and necrosis).4 Compared with these hyperenhanced area-size-assessed pre-ablation, the infarct-size induced by ASA was ~10-fold larger. In this respect, the induced myocardial infarct after septal ablation therapy enlarges the already existing arrhythmogenic substrate in HOCM patients. However, an electrophysiology report in high-risk patients after ASA has not indicated an increased arrhythmic substrate necessitating higher rates of implanting defibrillators.5 Although ventricular tachycardia and sudden death have been reported after ASA, these clinical features characterize the natural course hypertrophic cardiomyopathy irrespective of therapeutic LVOT gradient reduction. Further studies are necessary to evaluate the long-term effects of ASA with respect to ventricular arrhythmias and sudden cardiac death. Our goal in the near future must be developing additional tools to identify the high-risk HOCM patients, regardless of a potential intervention for LVOT obstruction, in whom defibrillator implantation is justified.

References

1. van Dockum WG, Kuijer JPA, Gotte MJW, ten Cate FJ, ten Berg JM, Beek AM, Twisk JWR, Hofman MM, Visser CA, van Rossum AC. Septal ablation in hypertrophic obstructive cardiomyopathy improves systolic myocardial function in the lateral (free) wall: a follow-up study using CMR tissue tagging and 3D strain analysis. Eur Heart J 2006;27:2833–2839.


Willem G. van Dockum
Department of Cardiology
VU University Medical Center
De Boelelaan 1117
PO Box 7057
1081 HV Amsterdam
The Netherlands
E-mail address: wg.vandockum@vumc.nl

Marco J.W. Gotte
Department of Cardiology
VU University Medical Center
De Boelelaan 1117
PO Box 7057
1081 HV Amsterdam
The Netherlands

Paul Knaapen
Department of Cardiology
VU University Medical Center
De Boelelaan 1117
PO Box 7057
1081 HV Amsterdam
The Netherlands

Albert C. van Rossum
Department of Cardiology
VU University Medical Center
De Boelelaan 1117
PO Box 7057
1081 HV Amsterdam
The Netherlands