

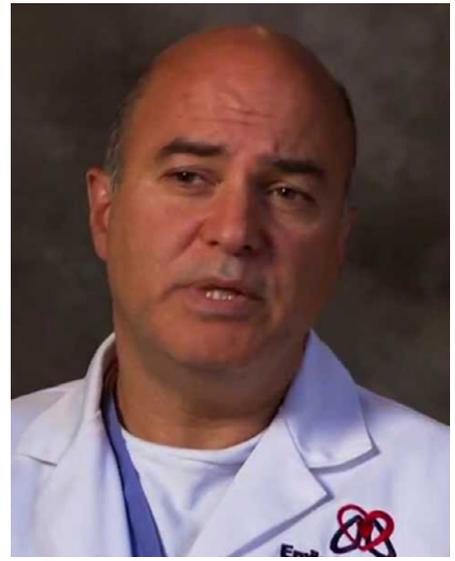
# Noonan's Syndrome and Cardiac Surgery: What Can We Learn?



Emile Bacha, MD, FACS<sup>\*,†</sup>

Noonan syndrome (NS), and autosomal dominant disorder, is the second most common genetic syndrome associated with congenital heart disease. Over 80% of patients with NS have cardiac involvement and the most common lesions are pulmonary stenosis, atrial septal defects, and hypertrophic cardiomyopathy (HCM). There clearly is a dearth of data with respect to outcomes after heart surgery for NS patients. The authors from the Mayo Clinic should be congratulated for putting together this valuable information spanning 16 years and including 29 patients. They describe their patient profile, give morbidity and mortality information, and elaborate on the nature of concomitant operations and their indications. We learn that NS patients do relatively well after cardiac repairs. Technical aspects of the operative approaches for left- and right-sided HCM are especially welcome since they come from a center with absolute preeminence in this area.

On the negative side of things, perhaps a more interesting study would have been to compare outcomes among patients undergoing similar operations with and without NS. This might provide more insight on to the importance of NS on outcomes after cardiac repair. However, the total number of patients is low and the spectrum of operations very wide so that propensity matching likely would not have been possible in a meaningful way. Half the patients were over 18 years old. Given the high redo-sternotomy rate, I assume many came to Mayo after having had surgery somewhere else. These facts may reduce the meaningfulness of the study to a more pediatric-oriented congenital heart center. Finally, the long-term follow-up, the Achilles Heel of most congenital heart studies, is frustratingly low. Nevertheless, the study will likely become a



Emile Bacha, MD, FACS.

### Central Message

Noonan's Syndrome patients often have congenital heart defects. A 17-year experience from the Mayo Clinic is described.

staple of the knowledge basis around NS and congenital heart surgery (especially for NS and HCM management), and serves to educate us all.

<sup>\*</sup>Division of Cardiac, Thoracic, and Vascular Surgery, New York-Presbyterian/Columbia University Medical Center, New York, New York

<sup>†</sup>Pediatric and Congenital Cardiac Surgery, Morgan Stanley Children's Hospital and Komansky Weill-Cornell, New York, New York

Address reprint requests to Emile Bacha, MD, FACS, Pediatric and Congenital Cardiac Surgery, Morgan Stanley Children's Hospital and Komansky Weill-Cornell, 3959 Broadway, CHN-274, New York, NY 10032. E-mail: [eb2709@cumc.columbia.edu](mailto:eb2709@cumc.columbia.edu)

DOI of original article: <http://dx.doi.org/10.1053/j.semtcvs.2018.12.004>.