The Journal of Thoracic and Cardiovascular Surgery

Elastin deficiency in Williams Syndrome may explain post-operative MACE

--Manuscript Draft--

Manuscript Number:	JTCVS-15-1303
Full Title:	Elastin deficiency in Williams Syndrome may explain post-operative MACE
Article Type:	Letter to the Editor
Section/Category:	CHD: Congenital Heart Disease
Manuscript Classifications:	20.1: Congenital AV valve repair/replacement; 20.2: Congenital aortic valve repair/replacement; 44: Pediatric perioperative management
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Elastin deficiency in Williams Syndrome may explain post-operative MACE

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Dear Dr. Weisel,

Dr. Christoph P. Hornik and co-authors established, in their paper you published in June 2015, that Major Adverse Cardiac Events (MACE) are common in patients with Williams syndrome (WS) following cardiac surgery. They suggest a multifactorial explanation.

WS is now recognised as a clinical manifestation of Elastin deficiency, with mutation sequenced in chromosome 7q11.23 [1]. Elastin, an extracellular matrix protein, is the major component of elastic fibres responsible for the elastic and rheological properties of arteries [2, 3]. Normal deposition of elastin is a critical event in vessel development [4]. Defective elastin has been implicated in aortic dissection for some time [5].

It would be therefore prudent to consider the pathological elastin as one of the factors that may explain the findings of Dr. Hornik.

Yours Sincerely

Aristotle D. Protopapas MSc. FRCS

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