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

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REFERENCES


