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LETTER TO THE EDITOR

Response to the letter, *Williams syndrome: was intubation rather than anaesthetic drug choice a cause of cardiac arrest?*

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Dear Editor

Response to the letter, *Williams syndrome: was intubation rather than anaesthetic drug choice a cause of cardiac arrest?*

We reported on the case of sudden death in a child with Williams syndrome following administration of anaesthesia,[1] and would like to respond to the letter to the editor by Cook,[2] which alluded to the fact that the death could most probably have been caused by intubation rather than the anaesthetic drug choice. The second author, Dr. JM Dippenaar, is a consultant anaesthesiologist, appointed as Clinical Head of Anaesthesiology in the Department of Maxillo-Facial and Oral Surgery at the Oral and Dental Hospital.

The fact that no evidence of myocardial ischaemia was found post mortem can be explained from a pathologist's perception as follows: Although early myocardial ischaemia is diagnosed fairly easily by clinicians with the use of an electrocardiogram, a pathological diagnosis is extremely difficult and depends upon the survival time of the individual after the infarction or ischaemia.[3] Usually, if the person survived for less than 12 h, macroscopic features will not be noted and the earliest findings will only be apparent from 30 min to 4 h after the ischaemia with histological examination.[4] These changes are very subtle and also depend upon the area of the heart sampled.

Intubation of this patient was necessary because of the severity of the dental caries. A dentectomy was envisaged; and, the dental surgeon preferred not to work with a supraglottic airway device, such as an *in situ* laryngeal mask airway (LMA). Furthermore, the appropriate size of a flexible LMA for this patient was not available in our institution. Mitigation of the stress response by means of additional agents, such as magnesium or a larger dose of opioid, was not considered because of the risk of hypotension.

The patient presented with hypotension and bradycardia, instead of the expected tachycardia and hypertension. This makes it unlikely that the sympathetic response after intubation was adequate to reverse the drug effects, let alone be of significance in facilitating the arrest, which was accompanied by severe ST segment depression, indicative of myocardial ischaemia. Decreased diastolic pressure in the presence of a physical obstruction to the right coronary was more likely to impact more severely on perfusion, especially in a hypertrophied ventricle where oxygen supply was already compromised.

The relationship between sudden death in patients with Williams syndrome and the administration of sedatives and anaesthetic drugs is very well documented;[5–8] and, includes most drugs used during sedation or anaesthesia, such as ketamine, propofol, sevoflurane, nitrous oxide (N_2O), halothane, pancuronium, diazepam, midazolam, choralhydrate, morphine, meperidine and promethazine.[9] It is questionable whether or not the use of muscle relaxation would have made any difference to this scenario since intubation was accomplished without any problem.

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