

## **GUIDELINES FOR GENERAL ANAESTHESIA – WILLIAMS SYNDROME**

### **1. Introduction:**

There have been very occasional cases of sudden death during general anaesthesia in children and adults with Williams Syndrome. These fatalities are rare and unpredictable, and generally relate to existing abnormalities in the cardiovascular system.

Nevertheless, we advise anyone parents/carers of anyone with WS who requires a general anaesthetic to read these guidelines, drawn up by consultant paediatrician Dr Neil Martin, a WSF Trustee and Chairman of our Professional Advisory Panel.

### **1. Children:**

General anaesthesia in children should only be undertaken in institutions with trained paediatric anaesthetists who have access to paediatric services.

### **2. Cardiovascular risks:**

Cases of sudden death have all been associated with significant cardiovascular anomalies – a common finding in WS. Where anaesthesia is performed for open heart surgery, these risks will be well recognised by the anaesthetist. Where surgery is for other problems, the anaesthetist may not be quite so aware of the potential problems. In view of these known associations, any individual with Williams Syndrome should have a pre-operative assessment including documentation of the cardiovascular status. This should have been performed within the last year and an ECG should be performed pre-operatively.

### **3. Hypertension:**

Up to 50% of adults with Williams Syndrome may have hypertension. Pre-operative blood pressures should be recorded and hypertension should be investigated prior to general anaesthesia.

### **4. Airway problems:**

Williams Syndrome may be associated with underdevelopment of the lower jaw and dental anomalies, including brittle or loose teeth. These should be identified in the pre-operative assessment.

### **5. Joint contractures:**

Likewise, due to the elastin disorder, some joints may have restricted movement. These contractures may lead to difficulties in positioning during anaesthesia.

### **6. Developmental delay/Emotional liability:**

Careful premedication may be necessary and avoidance of unnecessary distress is essential because WS people have a range of learning difficulties and some have features of Attention Deficit Hyperactivity Disorder or autism. They often appear loquacious and over friendly and may have considerable insight into their disorder. Many are frightened of loud noises and some may be needle phobic.



**7. Endocarditis prophylaxis:**

Bacterial endocarditis of the supravulval region has been reported but is rare. Routine antibiotic prophylaxis is no longer routinely advised. The 2008 NICE guidelines should be followed with respect to prophylaxis. These guidelines are currently under review and this advice may change.

**References:**

- a) NICE Guideline (CG64) – March 2008
- b) P Gupta et al. Annals of Cardiac Anaesthesia 2010:13; 44-48.
- c) T Burch et al. Anaesthesia and Analgesia 2008:107; 1848-1854