RED FLAGS ♥ BEST PRACTICES ♥ ANESTHESIA PRECAUTIONS

Heart Talk | Cardiovascular guidance for Williams syndrome

A candid, one-page reference on when to call and what to know before procedures.

RED FLAGS / WHEN TO CALL YOUR DOCTOR

- **Syncope or near-syncope** — passing out or nearly passing out.
  - Symptoms of passing out or near passing out
  - Passing out is not necessarily something very worrisome (most likely dehydration), but it needs to be checked out.
- **Symptoms with exertion** — Pain in leg, abdomen or other that increases with exertion and stops once recovered from exertion.
- **Change in exercise ability** — sustained decreased exercise tolerance, over a period of weeks to months.
  - This does not include during acute viral infection or other diagnosed illness.
- **Palpitations or a fast heart rate** — abnormal electrical heart activity that frequently has very abrupt increase, like a light switch on, not a ramping up like toaster heating up.
  - This heart rate increase is not explained by exertion, scary movie, fear cause by increased sympathetic activity (= increase in exertion, fear, anger, scary movie).
  - Can be tricky, as many people with Williams syndrome have an elevated baseline heart rate or sinus tachycardia.
  - Helpful question to ask: "Is there ever a time when you're sitting on the couch — not watching a scary movie — and your heart starts beating very fast for no reason?"
- **Gut feeling** — parents' intuition is often excellent, even when a specific problem can't be named.
  - Parents often have excellent intuition, even if they can't articulate a specific problem.
  - Even if you can't put your finger on a specific problem, but think something is not right, we want to hear from you.

WHAT YOU CAN DO NOW

- ✓ Keep a log of any symptoms; include exercise benchmarks if possible.
- ✓ Stay well-hydrated with small, frequent sips.
- ✓ Recommend conferring with the cardiac anesthesia team prior to any procedure.
- ✓ Continue regular annual visits to cardiology for exam, EKG, echo, Holter monitor, and labs to check BNP (brain natriuretic peptide) and creatinine.

ANESTHESIA PRECAUTIONS

1. Recommend anesthesia be provided only by those familiar with specific anesthesia risks in those with Williams Syndrome and only in a hospital setting.
2. Patient should only have anesthesia in hospital setting by individuals familiar with these risks and by those capable of managing patients with acute hemodynamic collapse in the unlikely event this occurs.
3. Recommend vigorous pre-hydration prior to any planned anesthesia administration.

When in doubt, call your cardiology team.

Developed in partnership between the Armellino Center of Excellence for Williams Syndrome and the Williams Syndrome Association.

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