

2. Cardiology

Monitoring cardiovascular health Aspirin for heart health



Monitoring cardiovascular health

Children with HGPS are at high risk for heart attacks and strokes at any age. Cardiovascular disease in Progeria is a gradual process. Blood pressure and ECG are often in the normal range until a child is older. Careful, repeat measurements are recommended because the best way to detect a problem is by asking if there has been a change over time.

The following testing should be considered annually, and more often if home physicians recommend:

- Cardiology visit with physical examination
- Measurement of fasting lipids and glucose
- Blood pressures of arms and legs
- Electrocardiogram (ECG)
- Echocardiogram
- Carotid duplex ultrasound*, if available
- Pulse wave velocity*, if available

Blood lipids such as cholesterol are very often normal in Progeria.

*Note, carotid duplex ultrasound and/or pulse wave velocity are available in some centers, but are not yet routinely performed on pediatric patients.

Low dose aspirin, dosed by weight at 2-3 mg per kg body weight, is often recommended.

Aspirin Treatments

Studies in adults have shown that the benefits of low dose aspirin therapy increase with increasing cardiovascular risk. Recommendations here come from experience in adults and in children with diseases which predispose them to heart attacks and stroke.

Low dose aspirin should be considered for all children with HGPS at any age, regardless of whether the child has exhibited overt cardiovascular abnormalities or abnormal lipid profiles. Low dose aspirin may help to prevent thrombotic events, including transient ischemic attacks (TIAs) stroke, and heart attacks, by inhibiting platelet aggregation. Aspirin dosage is determined by patient weight, and should be 2-3 mg/kg given once daily or every other day. Platelets may become “stickier” (i.e., more likely to form clots) at times of stress with illness, fever, etc. While these recommendations are guidelines, individuals may make adjustment in aspirin dosing based on their clinical course.

Once a child begins to develop signs or symptoms of vascular decline, such as hypertension, TIA, strokes, seizures, angina, dyspnea on exertion, ECG changes, echocardiogram changes, or heart attacks, a higher level of intervention is warranted. Antihypertensive medication, anticoagulants, anti-seizure, and other medications usually administered to adults with similar medical issues have been given to children with HGPS. All medication should be dosed according to weight, and carefully adjusted according to accompanying toxicity (negative side effects) and efficacy (effectiveness).

> Aspirin for heart health

Aspirin may rarely cause stomach discomfort. If excessive bleeding or bruising is detected, stop aspirin therapy and consult your physician. Aspirin therapy will probably need to be discontinued 1 week prior to any surgery; consult your physician if any surgery is being planned.

If your child becomes ill with chickenpox, stop the aspirin therapy (see below).

> Reye's Syndrome

There is a weak association between aspirin usage during Varicella (chickenpox) infection and Fatty Liver With Encephalopathy (Reye's Syndrome) in children under 15 years of age. The risk of Reye's syndrome is extremely small compared to the potential benefits of low dose aspirin treatment, given the risk of cardiovascular events in HGPS.