Important Information For You and Your Doctors About Low-Dose Aspirin Treatment and Progeria

Information for Families and Caregivers From The Progeria Research Foundation

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Our Goal

The Progeria Research Foundation hopes to improve health care of children with Hutchinson-Gilford Progeria Syndrome (HGPS) by providing medical and health recommendations to their families, physicians and caretakers. Because this syndrome is rare and there is little available literature from which to draw, there is often confusion and inconsistency in the care of these children. We hope that this treatment recommendation provides you with helpful information for use in current and future health care decisions for children with Progeria.

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Low-Dose Aspirin Treatment and Progeria

Why Low-Dose Aspirin in Children with Progeria?

Children with HGPS are at high risk for heart attacks and thrombotic strokes at any age. The earliest published incidence of stroke is at the age of 4 years[1] (1). In one case, seizures were the presenting cerebrovascular event[2] (2). Importantly, stroke (cerebral infarction) may occur while the child exhibits a normal EKG and may be due to occlusion of a small cerebral vessel in the absence of large-vessel intracranial blockages[3] (3).

Studies in adults have shown that the benefits of low dose aspirin therapy increase with increasing cardiovascular risk (4, 5). Our recommendations here are extrapolated from this evidence in adults. Given the risk of thrombotic events in children with Progeria, prevention of thrombosis should be a mainstay of treatment. *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 atherothrombotic events, including transient ischemic attacks (TIA) stroke and heart attacks, by inhibiting platelet aggregation.

What is the appropriate dosage and frequency in children with HGPS?

Dosage is determined by patient weight, and should be 3-5 mg/kg given once daily or every other day. This dosage will inhibit platelet aggregation but will not inhibit prostacyclin activity. It is usually quite well tolerated with low risks.

Warnings (reviewed in (4))

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 (chicken pox) 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 certainty of cardiovascular events in HGPS.

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[1] Smith et al. reported stroke in a boy who on MRA displayed bilateral occlusion of the internal carotid arteries, vertebral artery origins, and tortuous anterior spinal arteries along with subdural fluid collections indicating old hematomas.

[2] Rosman et al. (2001) presented a child who at 5 years of age developed left-sided clonic seizures. Angiography revealed a narrowed left internal carotid artery and occlusive disease of both cavernous carotid arteries.

[3] Wagle et al. (1992)) presented a child who at 8 years of age suffered his initial stroke due to occlusion of the superior division of the right middle cerebral aretry. MRI angiography using a 2D time-of-flight technique revealed normal patency of the large intracranial vessels (carotid artery bifurcations, vertebral and basilar arteries, and proximal middle cerebral arteries).

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