

HYPERCALCEMIA/HYPERCALCIURIA IN WILLIAMS SYNDROME



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WHAT IS CALCIUM AND WHY IS IT IMPORTANT?

Calcium is an important mineral for building teeth and bones and in chemical reactions, such as those that cause the heart to beat. It is found in high levels in dairy foods like milk and cheese, but also in some plant-based products like collard greens, tofu and white beans.

WHAT HAPPENS TO CALCIUM IN WILLIAMS SYNDROME?

People with Williams syndrome (also known as Williams Beuren syndrome or WS) may have problems handling calcium. Most children with WS have blood levels of calcium that are in the high normal range with a smaller fraction of affected individuals (around 15 percent) having very high levels of calcium (hypercalcemia). Hypercalcemia may require medications and/or changes in diet to bring the calcium level down. Children and adults with WS may also have high levels of calcium in their urine (hypercalciuria). People with hypercalciuria may or may not have high levels of calcium in the blood.

HOW WILL I KNOW IF MY CHILD HAS HIGH CALCIUM LEVELS?

Symptoms of high calcium in the blood include irritability, loss of appetite, vomiting and lethargy. Very high levels of calcium can cause seizures or problems with the rhythm of the heart. High levels of calcium in the urine can cause the calcium crystals to deposit in the kidney (called nephrocalcinosis) and may lead to kidney stones. Because of these findings, your doctor should check your child's calcium level with blood and urine tests. Because hypercalcemia tends to be a bigger problem in children than in adults with WS, the blood levels are generally checked more often in infants and young children.



HOW CAN I HELP PREVENT HIGH CALCIUM LEVELS IN MY CHILD?

Doctors have not yet determined exactly what causes the high calcium levels in children with WS (certain genes and regulatory units within the WS deletion have been suggested, but there is no definitive answer at this point). We know that the abnormal blood levels are most common in infants and small children, but cases of high blood calcium have been seen in adults on standard dose Vitamin D supplementation.

Because of the concern that Vitamin D may raise the level of calcium in the blood, current guidelines recommend that babies with WS not receive supplemental Vitamin D. However, breastfed babies with WS should be monitored by their doctors for signs of low Vitamin D and rickets. In specific cases, Vitamin D may be recommended.

Avoid feeding your child a diet that far exceeds the Dietary Reference Intake (DRI) of calcium for your child's age and size (see the table below for DRI values and for examples of high and low calcium foods). This may be a problem in children who have texture aversions and use large amounts of liquid supplements such as Pediasure to meet calorie goals, as these foods are often fortified with calcium.

Children over 1 year old should drink water daily. Water can help wash calcium crystals out of the kidneys and is generally good for the body.

RECOMMENDATIONS FOR ADEQUATE DIETARY CALCIUM INTAKE IN THE U. S.

AGE	CALCIUM INTAKE, mg/day
0 - 6 months	210
7 - 12 months	270
1 - 3 years	500
4 - 8 years	800
9 - 18 years	1300
19 - 50 years	1000
50+ years	1200

Source: US NAL

WHAT IF MY CHILD HAS HIGH CALCIUM LEVELS IN THE BLOOD OR URINE?

If your child has a small increase in the amount of calcium in the blood or urine, your doctor will likely recommend drinking more water or other fluids that do not contain calcium. Your doctor will also check to make sure your child is not taking in too much calcium for his or her age and size based on the DRI. A dietician or nutritionist may help you choose the most appropriate foods for your child (see the table of high and low-calcium foods below) as not taking in enough calcium can also cause problems.

HIGH AND LOW-CALCIUM FOODS

If blood levels of calcium become very high, your child may be referred to a specialist (most commonly a hormone doctor (endocrinologist) or kidney doctor (nephrologist)) to help manage the problem. Individual papers in the literature have offered different treatment options. In general, the doctors in our hospital favor giving fluid through an IV to lower very high calcium levels. In addition, they may use a type

HIGH CALCIUM FOODS	LOWER CALCIUM ALTERNATIVES
Milk, yogurt, ice cream, cheese	For infants: Calcilo XD specialty infant formula For children and adults: Rice Dream Classic Original or Rice Dream Classic Carob
Canned salmon (with bones), sardines, anchovies	Fresh (cooked) fish without bones, canned tuna fish
Tofu, white beans, chick peas, soy beans	Lima beans, black beans, green peas
Collard Greens, dandelion greens	Iceberg and romaine lettuces
Molasses	Maple syrup, dark corn syrup or honey (honey should not be given to children <1 yr old)
Calcium fortified foods such as orange juice, instant oatmeal, breakfast cereals, english muffins and other bread products with a longer shelf life	Unfortified juices and packaged goods, Examples: Tropicana Pure Premium Original orange juice, Mom’s Best Cereals (Oats and Honey Blend), Bob’s Red Mill Oats, fresh bread products baked from scratch

of medication used for osteoporosis called a bisphosphonate. A specialist will be needed to help choose the right treatment for your



child. Significantly elevated blood calcium is often transient and may not come back after treatment, but each child's course may be different.

High calcium in the urine tends to be a more long-term problem. A nephrologist is often involved in the management of hypercalciuria. Increasing fluids to help dilute and wash away the calcium crystals is generally recommended in all but the youngest children. Your doctor may also conduct an ultrasound of the kidneys to check for calcium deposits. In some situations, a medication may be given to change the pH of the urine, which may improve the body's ability to get rid of calcium.

ARE THERE PROBLEMS WITH LIMITING THE AMOUNT OF CALCIUM AND VITAMIN D MY CHILD WITH WILLIAMS SYNDROME GETS?

Long-term reduced intake of calcium and vitamin D are associated with weaker bones in adulthood (osteopenia) or, if the weakness is severe, osteoporosis. Weaker bones may put a person at higher risk of bone breaks in adulthood. In addition, not taking in enough calcium may actually cause urine calcium to increase.

Consequently, the practice of the Williams Syndrome Center at St. Louis Children's Hospital is not to limit calcium intake in children with WS and normal blood and urine calcium levels. In most cases, even children with mildly elevated calcium levels are not dropped below the DRI for age.

A NOTE ABOUT CALCIUM AND VITAMIN D IN TEENS AND ADULTS WITH WILLIAMS SYNDROME

Because of the risk of osteopenia in older individuals, it is recommended to monitor bone density in adults with WS. The current guidelines recommend a bone density scan (also called a DEXA) every five years starting at age 30. Vitamin D levels may also be tested to make sure that Vitamin D is not too low. The age at which this testing is begun is dependent on many factors, including whether the individual was ever on a calcium-restricted diet or whether the person has had bone breaks.

If low bone density or low vitamin D are noted, your family member with WS may be referred to an expert in bone and mineral diseases. If vitamin D supplementation is recommended, calcium levels should be monitored carefully (in some individuals calcium may go down due to the immediate effect of vitamin D on a hormone called parathyroid hormone (PTH), but rarely, the calcium level may become quite elevated). In general, high dose vitamin D supplementation is not recommended.

Weight-bearing exercise such as walking is also recommended to maintain cardiovascular and bone health. More strenuous exercise should be reviewed with the individual's cardiologist.

If you have additional questions about calcium problems in those with WS, please contact the Williams Syndrome Center at St. Louis Children's Hospital.

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Supported in part by a grant from HRSA H46MC24089



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