

Blood Draws / Questions & Answers for Families and Persons with PWS

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Q: What are some instances where my child would need blood work other than the genetic testing?

Children and adults with Prader-Willi syndrome, like any person with complicated medical conditions, may need to have blood tests done for a variety of reasons. It is important to remember that most blood tests look only for a single piece of information, such as a blood count, or to measure sodium or potassium in the blood. Therefore, although all the blood may be placed into a single tube, or just a few tubes, the sample is separated in the laboratory so that the technician that does each test ordered can have enough of the sample to complete the test and, if needed, repeat it to confirm the result. Each individual test is done using a different instrument or procedure. Therefore, more tests require more blood. Many blood tests have been developed to work on small amounts of blood, so that, on average, most blood draws can be less than two teaspoons of blood.

Children on human growth hormone therapy need to have regular monitoring of blood to be sure that the dose is correct. This testing measures insulin-like growth factor 1 (IGF1) which is part of the information the doctor needs to tell whether the dose of growth hormone is too low, too high, or just right. At the same time, the doctor may need to monitor the blood count or measure the output of the thyroid gland. Any time a person has surgery the doctor will likely request blood tests to confirm that the blood clotting factors are normal and that there are no unexpected abnormalities of electrolytes (salts and minerals in the blood) or blood counts that would increase the risk of problems during anesthesia or surgery. At the time of puberty, doctors may need to evaluate the body's hormone production to understand why outward signs of puberty are occurring earlier or later than expected. Blood tests may also be used to evaluate causes of seizures, to monitor drug therapies, to help identify the cause of infections or fevers, and to evaluate heart or kidney function.

It is important that you know what tests are being ordered for your child; therefore your doctor will be happy to explain the purpose for each of the tests ordered.

Q: Does the higher body fat content in a person with Prader-Willi syndrome contribute to the difficulty of blood draws? If so, why?

The process of drawing blood, also called "phlebotomy", is complicated, and in spite of how it sometimes looks, always difficult. Individuals with PWS tend to have extra fat in the front of the elbow, where it is safest and easiest to draw blood. This makes it more difficult to see or feel the vein, which makes it more difficult to get the needle into the vein. For this reason, some phlebotomists may prefer to draw blood from the back of the hand, where the veins may be somewhat easier to see, although they are usually smaller than the veins in the front of the elbow.

Like all things in life, some people are better at drawing blood than are others. If there have been problems drawing blood from your child in the past, don't hesitate to ask for the most experienced person for pediatric blood-drawing available. And, it is worth asking your doctor for recommendations about finding individuals with a lot of experience drawing blood from children. Often this may be a nurse who has worked in a neonatal intensive care unit or it may even be the doctor (or not!).

Q: I have been told that my child's veins are "like leather" causing the vein to roll when trying to get blood causing a lot of needle movement under the skin. Could you explain why or if the vein would roll and if it relates to PWS?

People who draw blood use a variety of terms to explain why they are having trouble getting the needle into the vein. In most cases the actual cause of the difficulty can't be determined. Extra fat in the area can make it harder to see the vein, but sometimes it makes it easier to insert the needle because the fat can support the vein and keep it

from moving. Sometimes the needle hits the vein and moves it instead of puncturing it. This is what people mean when they say the vein rolled.

It is possible that some people naturally have “tougher” connective tissue in the wall of the vein, making it more difficult to puncture, and therefore more likely to roll. Also, if a person has recently had blood drawn from a vein, or had an IV there, the vein may be visible, but have a blood clot in it that prevents the needle’s reaching the flowing blood.

Occasionally, if a person has had many blood draws from the same vein, the vein may become scarred and difficult to access. This is not common in children with PWS. Finally, veins have little flaps of tissue inside of them that act as one way valves to keep the blood flowing toward the heart. Sometimes the needle may enter the vein causing some blood to come out, and then, it stops flowing because the flap of the valve may cover the opening of the needle.

In general, children with PWS do better with blood draws, both in terms of the ease of success, and in terms of their behavior, than do other children, whether or not they have special needs.

Q: My child will not drink water, but the doctor says to hydrate her before getting blood drawn. She will drink milk. Is milk good enough to hydrate and how does drinking water/fluids help for a better blood draw?

When a person is dehydrated, there is less water in the body, and less in the blood. Therefore the veins tend to be somewhat constricted and harder to find. It is rare, though, for a child with PWS to be so dehydrated that it would interfere with a blood draw.

The reality is that the amount of water we need to take in each day is determined by what we call “resting energy expenditure”. This is a measure of how much energy the body needs each day, and it is largely determined by how much muscle (lean body mass) an individual has in his/her body. Since people with PWS have low lean body mass, their resting energy expenditure is less each day, which is why a person with PWS doesn’t need as many calories as another person of the same body weight. Since water needs are directly related to energy expenditure, a person with PWS also does not need as much water each day

as another person of the same weight. Our brains are well designed to tell us to drink when our body needs water. There is no evidence that this particular brain function is altered in PWS. Therefore, if a child with PWS seems to drink less than others it is likely because they actually need less. In my opinion, trying to force your child to drink more to increase the success of a blood draw is not likely to help, and is very likely to cause frustration in you and your child.

Milk is mostly water, and is therefore very good for maintaining hydration. The problem is that the 10% of skim milk that is not water is sugar and protein, both of which contain calories. Therefore, it would be unwise to use extra milk as a way to increase water intake, because it would also increase calorie intake. Your child should take enough milk to supply his/her daily calcium need, but should use water or calorie free flavored drinks for the bulk of the fluid intake.

Q: My son is fine with getting shots and the pinch of needles, but holding him still seems to upset him almost to hysterics - why?

Children with PWS are known to have increased tolerance to pain, and many parents note that their PWS child doesn’t seem to be bothered by needles for blood drawing or shots. For all children (and adults, for that matter) the anticipation of the needle is worse than the actual event. This is especially true for our PWS children, who have a natural tendency to “obsess” about upcoming events. Because young children don’t understand that they can’t move their arm when blood is being drawn, they usually must be held. If your child is able to be still, based on your past experience, let the phlebotomist know. Also, it may help if you offer to hold your child, because many children with PWS, like most children, don’t like to be restrained by strangers. Actually, neither do I.

Our children take many cues about how to behave from us. Therefore, if we appear to be frightened, upset or anxious about an upcoming blood draw our child will likely be frightened, upset or anxious. The most helpful thing that a parent can do is to remain calm and to be firm that having the blood draw is not a choice (having already asked the doctor what the blood test is for and why it is needed), and to not increase the child’s anxiety by making comments that are likely to increase it.

Q: My daughter gets bruised and pinched from the tubing each time she has blood work. Is there any way to make the rubber tourniquet used less painful and traumatic? Does it have to be "rubber-to-skin" contact to be effective?

As noted earlier, when the vein is more full with blood it is easier to get the needle in and get the blood out. The tourniquet (the rubber tubing) is meant to apply just enough pressure to the arm so that the blood in the artery is still entering the arm but that the blood in the vein, headed back toward the heart, can't get past the block. This is very similar to what happens when there is construction on the highway – cars can't get off, but more cars keep coming on and the road becomes very full.

The tubing does not need to be terribly tight to achieve this goal. The tourniquet itself should never cause bruising in a child with normal blood vessels and clotting ability. It is perfectly reasonable to apply the tourniquet over the shirt sleeve to prevent the rubber's touching the skin. Pinching sometimes occurs accidentally, and is difficult to completely avoid. In general, the more experienced the person who is drawing the blood, the less likely one is to have these problems. It is very difficult to draw blood without the use of the tourniquet. This is another of those situations that we just have to accept.

Q: My son was poked over 7 times, inside both elbows, tops of both hands, 1 ankle and his head was shaved to try that area. His veins kept collapsing and the nurse said they were "blown". What does that mean and why does that happen?

A vein is said to be "blown" when blood leaks out of the vein into the surrounding tissue. This is often accompanied by the rapid development of a clot to stop the blood from leaking. It is almost impossible to insert a needle into a "blown" vein because of the difficulty seeing or feeling the vein, and because of the clot. Sometimes when a needle is inserted into a small vein the body diverts blood away from the vein, so that blood can't be drawn out. This may be referred to as the vein "collapsing", but more often this is an explanation that is used for a vein that can be seen, but not accessed. That can occur for a variety of reasons.

The veins of babies are quite small, and drawing blood from a baby requires a special skill. While there is no standard of care, most providers feel that if they are not able to obtain the blood needed in two or three pokes, another phlebotomist should be called to try. In general, if blood cannot be obtained easily after an additional two or three tries, the doctor should be consulted to determine if the blood tests can be done at a later date. Sometimes, the tests can't be delayed, and the phlebotomist will begin to look for other sites from which to draw blood besides the usual veins in the front of the elbow (the "antecubital fossa" for those who like doctor-speak).

Whatever vein is used, there may be a small amount of leakage of blood into the surrounding tissue during the blood draw and when the needle is removed. The bruise is this blood that leaks into the tissue and remains there until the body can break down and remove the red blood cells. Bruises usually go from dark purple to blue to brown to yellow/green as the pigments in the red blood cells are gradually reabsorbed. Putting pressure over the puncture site for 2 to 5 minutes after the needle is removed can greatly reduce the bruising.

Blood can be drawn from any vein into which one can get a needle. The veins of the scalp are often easy to see in babies, and may occasionally be used, although it is difficult to get much blood out of these veins. They seem to work better for IVs. Scalp veins are rarely, if ever, used beyond a few months of age. Veins on the back of the hand, the top of the foot, the ankle, the groin, and occasionally in the neck may be used to draw blood, if needed. Rarely, the doctor may draw blood from an artery in the wrist or groin. Certain blood tests, for example blood-oxygen measurements, need to be done with arterial blood. Arterial blood should only be drawn by a person specially trained to do the procedure, usually a doctor, an intensive care nurse, or sometimes a respiratory therapist. The site of the puncture should be held with pressure for at least 5 to 10 minutes to prevent bruising.

Sometimes enough blood can be obtained for the necessary tests from a fingerstick, or heelstick in a baby. This procedure is most successful when the hand or foot is well warmed by wrapping it in a warm pack (a disposable diaper soaked with warm, but not too hot, water works very well). It is difficult to get more than 1 to 2 ml (1/4 to 1/2 teaspoon) of blood this way, so this approach may not always be adequate.

Using a warm compress may also help with drawing blood

from a vein, as does having a warm bath before the blood draw. When the skin gets warm the blood vessels dilate to help cool the body. This means there is more blood in the veins so it is easier to insert the needle.

Q. What is EMLA cream? Are there any other ways to numb the skin so the needle doesn't hurt?

EMLA stands for "eutectic mixture of local anesthetics". It is a combination of topical anesthetics blended into a cream that is applied to the skin, providing anesthesia at the surface of the skin. The cream takes 30 to 40 minutes to reach its full benefit, so it may need to be placed in multiple sites, in case there is trouble drawing blood from the first site tried. Care must be taken to see that the child doesn't accidentally eat the cream, and that the total dose applied is not excessive. Too much of these prescription medicines, whether they get in through the mouth or the skin, can have dangerous effects on the heartbeat. This is usually not a big problem, though.

Besides taking a long time to work, there are several other drawbacks to EMLA cream. First, it is expensive, and insurance doesn't always pay for it. Second, it only numbs the surface, and sometimes the more significant pain from a blood draw is caused when the needle moves around under the surface of the skin where it is not numb. Probably, and most importantly, the medicine also can cause the blood vessel to constrict, making it more difficult to get the needle into the vein.

There are other methods of numbing the skin, but most are either not effective or interfere with the blood draw, so they are less useful. EMLA is most useful for children who have severe anxiety before the blood draw. But, because most needle pokes actually are quick and hurt very little, the EMLA cream really doesn't do much. Children who are anxious, are likely to still be anxious, and the EMLA just makes them have to wait longer, and have more time to worry, before the blood draw. If your child has problems with anxiety about blood draws, and using EMLA

significantly reduces that anxiety, then it is safe and reasonable to use it. Unfortunately, it is fairly rare for it to make that much difference, especially in PWS children who generally find the waiting and the being restrained the worst parts of the blood draw.

Q: What is a "butterfly"?

A butterfly needle is specially designed with flat handles, or flaps, shaped something like a butterfly (you have to use your imagination) which make it easier for the phlebotomist to handle the needle. The flaps also make the needle more secure once it is in the vein, so it is less likely to be accidentally dislodged before the blood collection is complete. Butterfly needles come in very small sizes, so they are often the needle of choice for drawing blood from infants or children, or adults who have had difficult blood draws in the past.

Q: What can I do, as a parent, to minimize the trauma associated with blood draws?

It is important to always be an advocate for your child, and to work with your healthcare providers to ensure the best possible care. The most important thing the parent can do is to remain calm and collected, so that the child doesn't sense the parent's anxiety. Don't hesitate to ask questions about the purpose of the blood test. Also, sharing with the phlebotomist what has worked well for your child in the past (for example, distractions, being held by parent, not being held, putting the tourniquet over the shirt sleeve) can be a big help, and is usually better received than trying to tell them how to do their job. If you are becoming upset because the blood draw is difficult, suggest to the phlebotomist that you all take a short break. During that time you may be able to tactfully suggest that another provider be called to try, preferably one with even more experience. The phlebotomist will likely be thinking the same thing, but it may be easier for them to ask someone else if the request comes from you. Certainly talk to the doctor if you think that EMLA cream may help.

A special thank you to all the parents willing to share their experiences and questions concerning blood draws for their children and adults diagnosed with Prader-Willi syndrome.

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