

G6PD Deficiency

Glucose-6-phosphate dehydrogenase deficiency, or G6PD deficiency, is the most common enzyme deficiency worldwide.

Know more about it to protect your baby.

www.newbornscreening.ph

Newborn Screening Reference Center,

National Institutes of Health,

University of the Philippines Manila





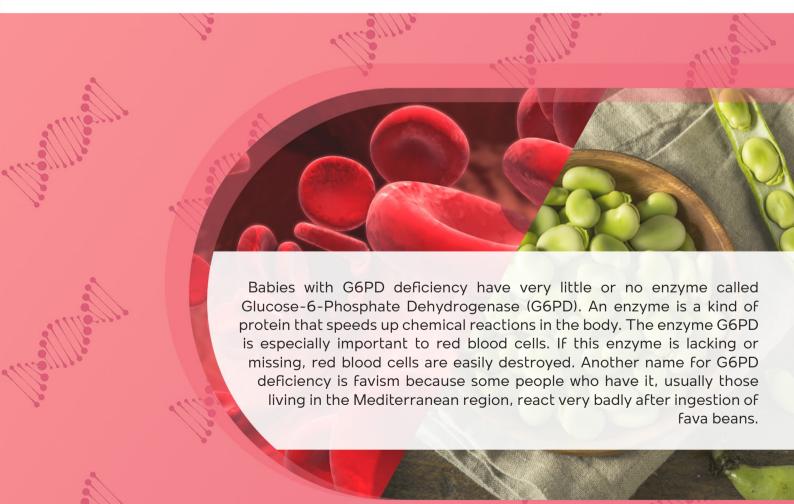




WHAT IS G6PD DEFICIENCY?

According to statistics, about 400 million people have G6PD deficiency, and it is most common in Africa, Southeast Asia and the Middle East. In the Philippines, data from the Newborn Screening Reference Center from 2004-2021 showed that 1 out of 63 newborn Filipinos have Glucose 6 Phosphate Dehydrogenase deficiency.

Glucose-6-phosphate dehydrogenase deficiency, or G6PD deficiency for short, is the most common enzyme deficiency worldwide. This is an x-linked inherited disorder which means that from the time a baby is born, there is already something wrong with how his body makes and breaks important substances.



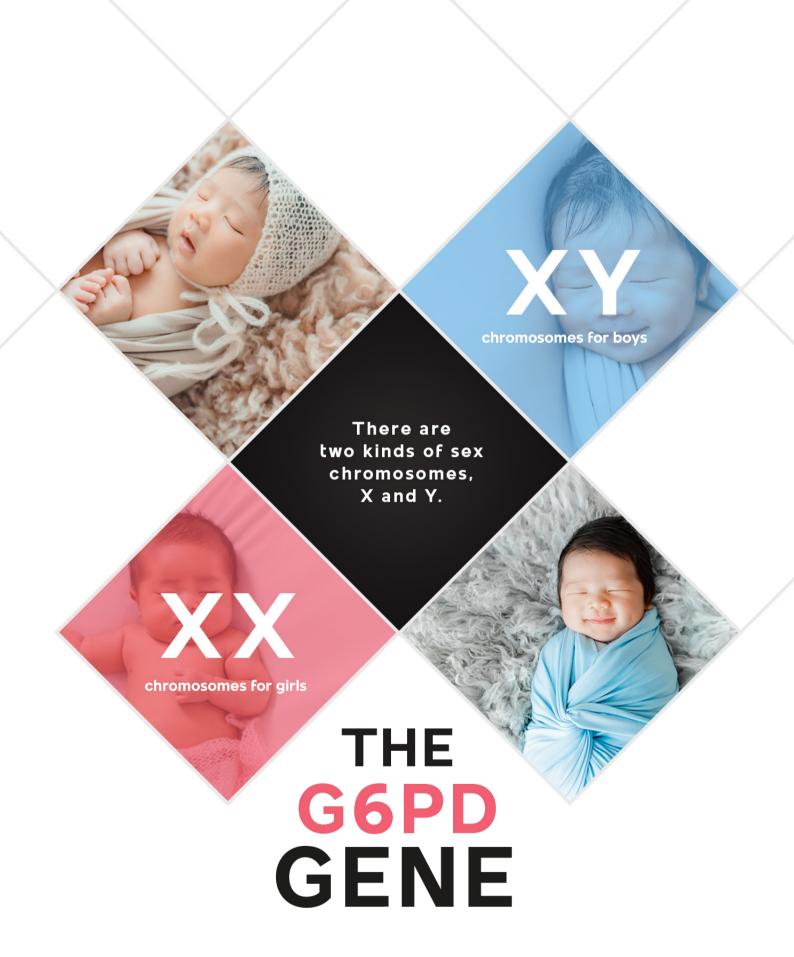
What causes G6PD deficiency?

In order to understand what causes G6PD deficiency, one must first learn a bit about genes and chromosomes. Genes are like the body's blueprints. They contain instructions on how specific parts of the body are made.

For example, if the instructions in your hair genes say your hair is black, your hair will be black. Genes are packaged into threadlike structures called chromosomes. A chromosome is very much like a beaded bracelet. The beads are the different genes that give instructions for different parts of the body; the entire bracelet is the chromosome.

Genes usually come and act in pairs. One member of a specific pair comes from the father, and the other member comes from the mother. The members of a pair are located on paired chromosomes.

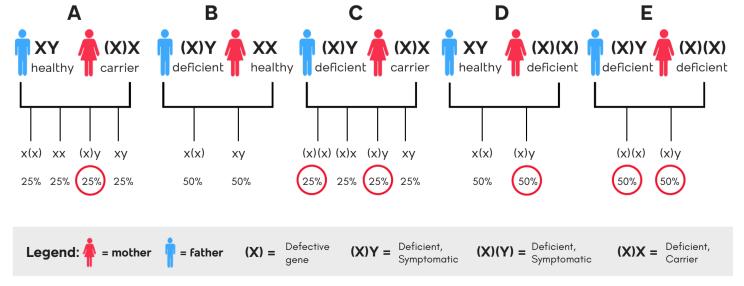
All normal human beings have 23 pairs of chromosomes. Each of the first 22 pairs contains the same number and kind of genes. The last and 23rd pair is the sex chromosomes. They are different from the first 22 pairs in that they do not have the same number and kind of genes. The sex chromosomes contain the genes that determine whether a baby will be a girl or a boy.

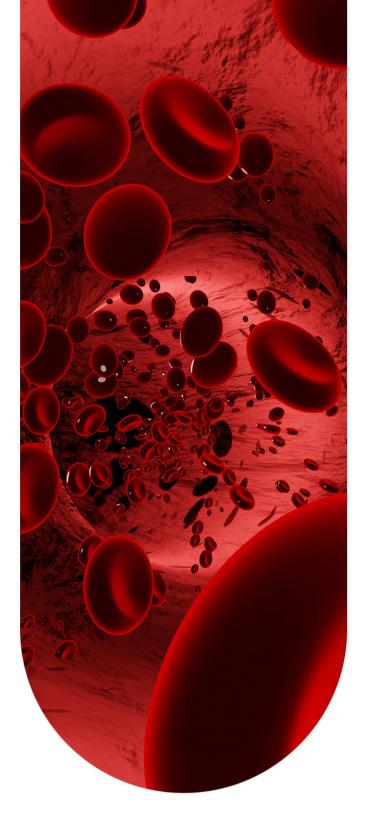


All baby girls have two X chromosomes. All baby boys have one X and one Y. The gene that gives instructions on how G6PD is made is found in the X chromosome only, thus G6PD deficiency is described as X-linked.



Inheritance of G6PD Deficiency





G6PD has a very small but strategic role in protecting the body from substances that can cause damage to cells or oxidative substances. Because of this important role, G6PD is normally found in all parts of the body. To be sure, most parts of the body also keep a "spare" enzyme, one that can do the work of G6PD in case it is lacking or missing entirely. Unfortunately, this is not the case with red blood cells. They do not have spare enzymes that can do the work of G6PD. If a baby does not have enough G6PD, his red blood cells lack protection from the harmful effects of oxidative substances.

What are the RISKS FOR AN INDIVIDUAL with G6PD Deficiency

A baby with G6PD deficiency appears and remains healthy until he is exposed to a large amount of oxidative substances. When this happens, his red blood cells are destroyed, a process known as hemolysis. Red blood cells carry oxygen to all parts of the body. When they undergo hemolysis, the baby will have hemolytic anemia. The signs and symptoms of hemolytic anemia are paleness, dizziness, headache, tea-colored urine, and abdominal or back pain or both. Hemolytic anemia, when very severe, can end in death.

Destroyed red blood cells are brought to the liver to be broken down to smaller pieces for disposal. One of the end products of this process is bilirubin, a yellowish substance that accumulates in different parts of the body when too much of it is produced. Quite often, bilirubin accumulates in the skin and causes it to appear yellowish. In the worst cases, bilirubin accumulates in the brain and causes mental retardation or death.

WHERE DO OXIDATIVE SUBSTANCES COME FROM?

Hemolysis of red blood cells will only occur IF and WHEN a G6PD deficient child is exposed to oxidative substances. Oxidative substances are found in certain drugs, foods, and beverages. The body also produces oxidative substances during severe infections or illnesses such as typhoid fever, pneumonia, or kidney failure. **Most drugs with strong oxidative effects are of these kinds:**



1. antibiotics of the sulfa group



2. medicines for malaria



3. some medicines for fever

For a more detailed list of drugs, food, and beverages with strong oxidative substances, please refer to page 9 and 10.

How is G6PD deficiency treated?

When a child has taken oxidative substances and suddenly shows the signs and symptoms of hemolytic anemia, he is said to have a hemolytic crisis. During such crisis, the goal of doctors and nurses is to prevent the harmful effects from getting worse. Blood transfusion, oxygen, and folic acid may be given. The ultimate treatment for G6PD deficiency is gene therapy (replacing a defective gene with a good one), but this is not yet available at the present time.



As a parent, how do I prevent a HEMOLYTIC CRISIS?

Tell your child's pediatrician that your child has G6PD deficiency. This is very important so that he will not prescribe oxidative drugs in case your child gets ill. He would also be able to watch out for hemolytic crisis and would immediately know what to do just in case it happens.

Keep your list of oxidative substances in a handy place. Better yet, post it in a convenient spot on the kitchen wall. Always double-check food, beverage, & medicine labels against the list.

Memorize the signs and symptoms of hemolytic anemia: paleness, dizziness, headache, difficulty in breathing, rapid and strong heartbeats, tea-colored urine, and abdominal or back pain. Bring your child to his pediatrician as soon as these signs and symptoms appear.

My baby did not go through newborn screening. What should I do to make sure that he/she is not G6PD deficient?

You can bring your child to a pediatrician for a thorough medical check up. If the age of your baby is already beyond the limit for newborn screening, he/she can still undergo confirmatory testing for G6PD deficiency. However, for the other diseases included in the newborn screening, it will depend on the assessment of your pediatrician if your baby will need a referral to a specialist (i.e. endocrinologist, metabolic geneticist, hematologist, pulmonologist) for further evaluation.

Do not ignore infections. Persistent fever signals an infection. Bring the child at once to his pediatrician.

As your child gets older, honestly and gently tell him about his condition and teach him to be careful about what he eats.



My baby has G6PD deficiency and I feel devastated. Did I do something wrong when I was pregnant? Could I have done something while I was pregnant for my baby to avoid it?

You did nothing wrong. It is a genetic disorder, hence, there is nothing you can do as a mother to prevent it.

My firstborn was diagnosed with G6PD deficiency. I am pregnant again and worried — will my next baby also have it?

Once you have a child with G6PD deficiency, there is always a possibility of having a child with G6PD deficiency for every succeeding pregnancies. All your babies must undergo newborn screening.

Even if tested positive by Newborn Screening Test, is it possible that the confirmatory test of my child is negative?

Yes. If negative, then you do not have to worry about the medicines and the food items. If positive, make sure that your attending physician knows that your child has G6PD deficiency.

I am a parent. Should I get tested for G6PD deficiency?

Yes, you may want to have yourself tested by requesting for G6PD assay using the confirmatory test in designated confirmatory centers. However, only males who are G6PD deficient and affected females (homozygous for G6PD) can be identified. A female with a carrier status (heterozygous for G6PD) cannot be detected by this kind of test.

I am trying to conceive. Should I be tested for G6PD deficiency?

It is your choice if you want to be tested but the probability of having a child with G6PD deficiency does not solely depend on your status. Your spouse's G6PD status can also affect the probability of having a child with G6PD. It will be more practical to just have your newborn baby undergo the routine newborn screening after birth.

Where is G6PD Deficiency Confirmatory Testing done?

At the moment, there are 31 Confirmatory Testing

Centers in the country. You may want to contact them directly regarding their testing schedule. Please visit www.newbornscreening.ph for details.

Is formula milk safe for my son who has G6PD deficiency?

Although soya and soy lecithin are both included in the list, many products contain very small amount of soya including the common milk formulas. There has not been any report of hemolysis due to milk products containing soya. Soy lecithin, which is present in different brands, is an important component of milk but it does not make up the whole milk. REMEMBER: Breastfeeding is still best for babies. Breastfeeding is healthier for both babies and mothers.

I am breastfeeding my son. When I haven't received the newborn screening result I ate the food that need to be avoided. Will they take effect on my son?

Theoretically, food and chemicals can be excreted through the milk. If your son did not develop hemolysis due to that event in the past, then probably they were not excreted at all through the milk or the amount was too small to cause significant reactions.

I have donated blood several times without knowing I am a G6PD carrier. Can I still donate blood?

Yes, you may still do nate. There is no contrain dication as a person with G6PD deficiency. There is also no policy written that you cannot do nate.

Is it transmissible via blood transfusion?

No, it is not transmissible via blood transfusion.

My baby has G6PD deficiency; vitamins are on the restricted list. How can I keep him healthy?

Multivitamins are generally not contraindicated. Vitamin C or ascorbic acid can be given for as long as the dose is within the recommended dosage. Multivitamins are considered food supplements, BUT there is no substitute to nutritious, freshly prepared food and a well-balanced diet.

Is it okay to eat chocolates even if they have soy content?

Although soya is included in the list, many products contain very small amount of soya. So generally patients with G6PD deficiency can eat chocolates except if it is a fava bean coated with chocolates. Please read the product label.

Is malunggay leaves safe to eat? Because I read in some website that malunggay isn't safe for people with G6PD deficiency.

For as long as it is not included in the official list of NIH as foods to avoid, then there is no basis to withhold malunggay from persons with G6PD deficiency.

I have a son who has G6PD deficiency and now he has a cold and cough, it is safe to use oregano for his medication?

Since oregano is not included in the list, it can be given. However, you should exercise caution in giving commercially produced herbal preparations because they might contain unreported chemicals. Generally, cough and colds can be treated with lots of fluids. If signs and symptoms persist or worsen, consult your doctor.

Is infant cereal safe for newborns with G6PD deficiency?

So far, there are no reported cases of hemolysis secondary to rice infant cereals (eg Cerelac). It contains a very small amount of soya in the first place.

Is insect repellant safe to use?

If you are not comfortable in using insect repellant, you can look for an organic type. If not available, you can just apply the commercial ones on your baby's clothes.

What toothpaste brand is safe for babies with G6PD?

The amount of menthol that is in ordinary toothpastes so far has not caused hemolysis in patients with G6PD deficiency.

What about blueberries? In other countries, they are not listed as contraindicated food.

If there is confusion about blueberries then it might be best to avoid it until extensive evidence that it is safe for patients with G6PD deficiency becomes available. Please note that patients with G6PD deficiency are not the same all over the world because of differences in mutations and therefore phenotypes, some of them will react consistently with some of the items in the list and some would not.

What are the signs of hemolysis?

Signs of hemolysis include tea colored urine, discoloration of the eyes and skin, weakness, and irritability. A child may look clinically pale but may actually have normal CBC results. So a CBC should be requested to confirm if the child has anemia.

My son exhibited symptoms of a viral infection (vomiting, lbm, lack of appetite). I saw on his diaper that the stain of his urine is somewhat tea colored. Should I be worried? What should I do?

Bring him to his pediatrician for check up. Please do CBC APC, and urinalysis. Check for urine hemoglobin.

Have there been reported cases of hemolysis here in the Philippines because of G6PD deficiency? If so, what were the most common causes?

Yes, there have been reports of hemolysis among patients with G6PD deficiency. There are also claims registered at PhilHealth. Some of the causes of hemolysis recorded in Philippine Children's Medical Center are infections (Flu, hepatitis) and exposure to moth balls. Fortunately, none of these patients died because of severe anemia. They were brought immediately to the hospital as soon as the initial signs of hemolysis became evident

What are the chances of getting a different result if we undergo another Confirmatory Test?

The family of a child confirmed with G6PD deficiency may always opt to have another confirmatory test if they are having doubts. If the result is below the borderline, the patient will most likely get a positive result in another test.

As a mom, can I use menthol-containing products?

The use of menthol and camphor containing products are not recommended if the goal is to reduce or eliminate the pain. These products only divert attention away from the painful area by producing cooling effects and sensation on the skin. It is always better to know the real cause/s of the pain/discomfort by consulting your doctor about it.

Is there a contraindication to any vaccination for patients with G6PD deficiency?

None.



DRUGS TO BE AVOIDED

As of August 2023

A. Antibacterial

**Nalidixic acid* Nitrofuran

- 1. Nitrofurantoin
- 2. Furazolidone
- 3. Nitrofurazone / nitrofural

(Common Brand Names: Macrodantin Diafuran, Diapectolin, Furoxone, Furacin)

*P- aminosalicylic acid

B. Analgesic/ Antipyretic

*Acetanilid

C. Antihelmentic

- *B-naphthol
- * Niridazole
- *Stibophan

D. Sulfonamides and Sulphones

Dapsone (Common Brand Name: Lepravir)

*Glucosulphone sodium

Glyburide/Glibenclamide (Common Brand Name: Euglucon, Gluban, Lodulce, Orabetic)

*Mafenide acetate

*Salicylazosulphapyridine/Sulfasalazine

Stibophen (Common Brand Name:

2-(2-Oxido-3,5-Disulphonatophenoxy)-1,3,2, Benzodioxastibole-4-6-Disulphonate)

*Sulphadimidine

*Sulphafurazone

Sulphamethazole/Sulfamethazole

(Common Brand Name: Bacidal, Bactille Forte, Bactrim, Bacxal, DLI Cotrimoxazole, Forteprim, Globaxol, Pharex Cotrimoxazole, Ritemed

Cotrimoxazole, Septrin, Trim S)

Sulphanilamide/Sulfanilamide

Sulphapyridine

*Sulphoxone/Sulfoxone

Sulfasalazine, Salazosulfapyridine

(Common Brand Name: Salazopyrin)

E. Antimalarials

Chloroquine

(Common Brand Name: Aralen, Chlorofoz)

*Pamaquine

Primaquine

Pentaquine

F. Miscellaneous

Acetylphenylhydrazine

Dimercaprol

Futamide

Isobutyl nitrate

Mepacrine

Phenazopyridine (Common Brand Name: Azomir)

Probenecid

Thiazolesulfone

Urate oxidase/Rasburicase

You may visit www.newbornscreening.ph to check for updates on these lists

DRUGS SAFE TO TAKE IN THERAPEUTIC DOSES

Acetaminophen (Paracetamol, tylenol)

Acetophenetidin/phenacin

Aspirin/ Acetylsalicylic acid (Alka-seltzer, Aspilets, Cor-80, Cortal)

Ascorbic acid

Chloramphenicol (Chlormycetin, Chloro-S, Chlorsig, Klorfen,

Oliphenicol, Optomycin, Pediachlor, Penachlor Speradex)

Ciprofloxacin (Ciprobay, Clpromax, Cipromet, Qinosyn-500, Quilox, Xipro)

Diphenhydramine

Isoniazid

Phenytoin

Quinidine

**Vitamin K analogues/Phytomenadione

(Hema-K, Konakion MM, Phil Pharmawealth/Atlantic Phytomenadione)

NOTE:

*Not available in the Philippines

**Should be water soluble

IMPORTANT REMINDERS FOR PATIENTS WITH G6PD DEFICIENCY

- 1. Avoid ingestion of or exposure to the listed drugs and chemicals.
- 2. If you have cough, colds, or other bacterial or viral infections, make sure to inform your doctor that you have G6PD deficiency.
- 3. If you have ingested or were exposed to any medication and your urine became tea-colored, inform your doctor immediately.
- 4. If you have yellowish discoloration of skin, sclera or any part of your body, consult your doctor immediately.

CHEMICALS

- - Methylene Blue
- Arsine
- Phenylhydrazine
- Toluidine blue Trinitrotoluene
- Aniline dyes

FOOD AND DRINKS

- Fava beans (Dingdong nuts, Mr. Bean)
- Red wine
- Legumes (Abitsuelas, Garbanzos, Kadyos, Munggo)
- Blueberry
- Soya food (Taho, Tokwa, Soy Sauce)
- Tonic water
- Bitter melon/ampalaya

OTHERS

- Menthol (Alaxan Gel, Ben-gay, Efficascent Oil, Listerine Mouthwash, Listerine Pocketpacks Megascent Oil, Mentopas Medicated, Plaster, Omega Pain Killer)
- Camphor (Liniments)
- Naphthalene (Moth balls)
- Parabenzene dichloride / dichlorobenzene (Toilet deodorizer)
- Henna
- Herbs (Cattle gallstone bezoar, Honeysuckle flower, Chimonanathus flower, 100% pearl powder

Figwortflower, Acalypha indica)

G6PD Confirmatory Centers



As of Aug 2023

			As of Aug 2023
	CENTER	ADDRESS	CONTACT
CAR			
1	Baguio General Hospital and Medical Center	Gov Pack Rd., 2600 Baguio City	09332003296
REGION	1		
2	llocos Training and Regional Medical Center	Parian, San Fernando City, La Union	09778108778
3	Mariano Marcos Memorial Hospital & Medical Center	Brgy. 6, San Julian City of Batac, Ilocos Norte	(077) 600 8000
4	Region 1 Medical Center	Arellano Street, Poblacion Oeste Dagupan City	(075) 515-8916
REGION	2		
5	Cagayan Valley Medical Center	#2 Dalan na Pagayaya Carig Sur, Tuguegarao City	(078) 302 0000
REGION	3		
6	Angeles University Foundation Medical Center	Mac Arthur Hi-way, Angeles City	09562943730
7	Bataan General Hospital and Medical Center	Manahan St., Tenejero, Balanga City	(047) 237-1275 loc 6704
REGION 4	I-A		
8	Lipa Medix Medical Center	J.P. Laurel Highway, Balintawak, Lipa City	09338526522; 09611191788

National Highway, Sto. Niño,

Biñan City, Laguna

(02) 8779 5310;

(049) 544 5150 loc 2033

9

University of Perpetual

Help Dr. Jose G. Tamayo

Medical Center- Binan (Perpetual Help Medical

Center-Biñan)

G6PD Confirmatory Centers



			As of Aug 2023		
	CENTER	ADDRESS	CONTACT		
10	St. Anne General Hospital	P. Gomez Ext., Red-V, Ibabang Dupay, Lucena City, Quezon	09228985922; (042) 7103506; 7102218		
REGION 4	I-B				
11	Palawan Medical Mission Group Multipurpose Cooperative	Corner Burgos & Mabini Sts., Puerto Princesa City, Palawan	09989565102		
NCR					
12	Manila Central University- Dr. Filemon D. Tanchoco Sr. Medical Foundation, Inc.(MCU-FDTMF, INC.)	Samson Road, EDSA, Caloocan City	(02) 8363 3084		
13	Our Lady of Lourdes Hospital	46 P. Sanchez St., Sta Mesa Manila	(02) 8716 3901 loc 3399/3396/3397; 09218528516		
14	The Medical City	Ortigas Avenue, Pasig City, Metro Manila	(02) 8988 1000 loc 6125; 8635-6789 loc 6125		
15	University of Perpetual Help Dalta Medical Center	#2 Dalan na Pagayaya Carig Sur, Tuguegarao City	(02) 8874 8515 loc 153; 88748515 loc 151		
REGION 5					
16	Bicol Regional Training and Teaching Hospital	Rizal St. Legazpi City Albay	(052) 483 1088; 483 0806		
17	Bicol Region General Hospital and Geriatric Medical Center	San Pedro, Cabusao, Camarines Sur	09637045263		
REGION 6					
18	West Visayas State University Medical Center	E. Lopez St., Jaro, Iloilo City	(033) 320 2431 loc 108/138		
19	Dr. Pablo O. Torre Memorial Hospital	B.S. Aquino Drive, Bacolod City 6100, Negros Occidental	09988460620		

G6PD Confirmatory Centers



As of Aug 2023

			J
	CENTER	ADDRESS	CONTAC
REGIO	N 7		
20	Cebu Doctors' University Hospital	Osmeria Boulevard Capitol Site, Cebu City 6000	(032) 255 5555
21	Gov. Celestino Gallares Memorial Hospital	M. Parras St., Tagbilaran City, Bohol	(038) 411 4868; 411 4869
22	Silliman University Medical Center	V. Aldecoa, Daro, Dumaguete City, 6200	09453902877
REGIO	N 8		
23	Eastern VIsayas Regional Medical Center	Brgy. Bacagay, Tacloban City	(053) 833 5309; 09771178444
REGIO	N 9		
24	Brent Hospital	R.T. Lim Blvd. Zamboanga City	09953273022
REGION	N 10		
25	Cagayan De Oro Polymedic Medical Plaza	Kauswagan Highway, Cagayan De Oro City	09173051163, 09688523615
26	La Vina General Hospital	L. Alkulro St. Valencia City, Bukidnon	09976058050
27	Mayor Hilarion A. Ram I ro Sr. Medical Center	Maningcol, Ozamiz City	(088) 521 0440
REGIO	N 11		
28	Tagum Doctors Hospital	Highway SA Rabe Subd., Tagum City	09195451830
29	Davao Medical School Foundation Hospital	DMSF Drive, Bajada, Davao City	09266887834
REGION	N 12		
30	General Santos Doctors Hospital	National Highway, General Santos City	09338162793

Quezon Blvd. Kidapawan City

09237234746

31

Kidapawan Doctors

Hospital

G6PD

INFORMATION ABOUT THE GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY

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