

Recommendations for Management of Duarte Galactosemia (D/G)

Dear Doctor,

Duarte Galactosemia is a benign variant of classic galactosemia. Infants with D/G galactosemia usually have a galactose -1- phosphate uridylyltransferase (GALT) activity between 5 and 20% of control values. These infants have inherited one gene for classic galactosemia from one parent and one gene for the Duarte variant from the other parent. There is a short video available on our website which provides additional explanation of Duarte galactosemia (www.peds.ufl.edu/peds2/divisions/genetics/caw/programs_newborn_screening.htm).

There is no strong evidence that suggests there are any complications associated with D/G galactosemia. There is disagreement in the metabolic community about the need for dietary restriction of galactose for these infants. Given that there is no clear evidence that galactose restriction is beneficial, we have developed the following recommendations:

1. **For Breast-fed Babies:** Continue breastfeeding and check the RBC Galactose-1-Phosphate (Gal-1-P) levels monthly. If the Gal-1-P level is above the normal range, continue breastfeeding but consider substituting 1-2 feedings per day with a powdered soy-based formula, such as Prosobee, Isomil or Good Start Soy. Recheck the Gal-1-P level every 2-6 weeks and make diet adjustments thereafter.
2. **For Formula-fed Babies:** Feed soy-based formula, such as Isomil, Prosobee or Good Start Soy, and restrict dairy products until the infant is 1 year old. At one year, we recommend gradually adding dairy products over a one month period and then checking the Gal-1-P level to make sure that it is not elevated. A protocol for the introduction of galactose is available.
3. Blood for Galactose-1-Phosphate, RBC levels (Gal-1-P) should be collected and sent to either Mayo Medical Laboratories or Emory Biochemical Genetics Program. Addresses are:

Mayo Medical laboratories
3050 Superior Drive NW
Rochester, MN 55901
1-800-533-1710

Emory Diagnostic Laboratory
Dept. of Genetics and Molecular Medicine
Emory Clinic, Rm. 412, 1262 Clifton Road
Atlanta, GA 30322
(800) 727-8308 FAX (404) 727-8310

4. Genetic counseling is advised for families who have babies with D/G genotype. If the family desires genetic counseling, please call the Division of Genetics to make a referral.

We are happy to discuss treatment recommendations with you or with your patient's family. Our clinical physicians and our nutritionists are available to provide consultation. We can be reached at 352 392-4104 or through the Shands Consult Center at 1-800-749-7424.

Thank you,

Charles A. Williams, MD
Professor
Division of Pediatric Genetics