



Newborn Screening Quality Assurance Program

PROFICIENCY TESTING

Sickle Cell Disease and Other Hemoglobinopathies

Volume 21, No. 1

Panel 1

February 2011

INTRODUCTION

On January 18, 2011, we distributed five dried-blood-spot (DBS) specimens prepared from umbilical cord bloods to all active participants for the Panel 1 Sickle Cell Disease and Hemoglobinopathies Proficiency Testing (PT) event. A total of 77 panels were mailed by overnight FedEx mail. The packages went to 50 domestic laboratories and 27 foreign laboratories. This PT report is a compilation of data reports received from 73 of the participating laboratories by the designated deadline date. There were 4 laboratories that did not report this quarter. We distribute this quarterly report to all participants, state laboratory directors, and to program colleagues by request.

We requested that participants assay all survey specimens by the analytical schemes they routinely use and report for each specimen the presumptive phenotype, the presumptive clinical assessment, and any other clinical classifications that they deem consistent with their analytic results and program operations. ❖

PARTICIPANTS' RESULTS

The certification report listing hemoglobins (Hb) by phenotype and their presumptive clinical assessments appears on page 2.

The frequency distribution of reported presumptive phenotypes and clinical assessments appears on page 3.

The individual data verification for each laboratory with evaluation comments appears on page 4.

We will continue to ship three PT panels this year for Hemoglobinopathies, therefore, the next shipment for the Hemoglobinopathy PT program will be on May 2, 2011. ❖

MEETINGS AND TRAINING

Hemoglobin Disorders Training Course: Laboratory Diagnosis and Clinical Management, April 10, 2011 - Brussels, Belgium

SCDAA 39th Annual Convention, September 27, 2011 - October 1, 2011, Memphis, TN

SPOTLIGHT

A new compound appears to prevent the traffic jam of cells that causes debilitating pain crises and associated mortality in sickle cell disease, Medical College of Georgia researchers report. An oligonucleic acid or peptide molecule that binds to specific target molecules was developed by the Archemix Corporation in Cambridge, Mass. This aptamer appears to work by occupying sticky receptors lining the walls of small blood vessels where sickle-shaped red blood cells and white blood cells can pile up, according to the study published by Diana R. Gutsaeva, James B. Parkerson, Shobha D. Yerigenahally, Jeffrey C. Kurz, Robert G. Schaub, Tohru Ikuta, and C. Alvin Head, Inhibition of cell adhesion by anti-P-selectin aptamer: a new potential therapeutic agent for sickle cell disease, *Blood*, Jan 2011; 117: 727 - 735. 13 January 2011, Vol. 117, No. 2, pp. 727-735.

ACKNOWLEDGMENTS

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CDC/APHL

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**Newborn Screening Quality Assurance Program
Sickle Cell Disease and Other Hemoglobinopathies**

Specimen and Lab Certification

Year: 2011 Panel: 1

Presumptive Clinical Phenotypes

	Specimen 11H1	Specimen 11H2	Specimen 11H3	Specimen 11H4	Specimen 11H5
Expected Presumptive Phenotype	FA	FAS	FAC	FA	FA
Accepted Presumptive Phenotypes	FA + Fast bands	FAS	FAC	FA + Fast bands	FA + Fast bands

Presumptive Clinical Assessments

	Specimen 11H1	Specimen 11H2	Specimen 11H3	Specimen 11H4	Specimen 11H5
Expected Presumptive Clinical Assessment	01	02	03	01	01
Accepted Presumptive Clinical Assessments	01, 22	02	03	01, 22	01, 22

- 01 Normal--no abnormal Hb found
- 02 Hemoglobin S carrier
- 03 Hemoglobin C carrier
- 04 Hemoglobin SS disease (Sickle cell anemia)
- 05 Hemoglobin SC disease
- 06 Hemoglobin SD disease
- 08 Hemoglobin D carrier
- 09 Hemoglobin E carrier
- 12 Hemoglobin SE disease

- 16 Alpha-thalassemia (Bart's Hb)
- 18 Hemoglobin EE disease
- 20 Assessment not listed
- 21 Unsatisfactory specimen
- 22 Unidentified variant, fast or aging band
Specimen not evaluated (NE)

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Frequency Distributions

Year: 2011 Panel:1

Phenotypes			Clinical Assessments		
Specimen Number	Hemoglobin Phenotypes	Frequency Distributions	Specimen Number	Presumptive Assessments	Frequency Distributions
11H1	FA	71	11H1	01 Normal	72
	FF	1*		22 Unidentified variant, fast or aging bands.	1
	FAV	1*			
11H2	FAS	71	11H2	02 Hemoglobin S carrier	72
	FSA	1		04 Hemoglobin SS Disease	1*
	FS	1*			
11H3	FAC	66	11H3	03 Hemoglobin C carrier	73
	FCA	7			
11H4	FA	72	11H4	01 Normal	73
	FF	1*			
11H5	FA	73	11H5	01 Normal	73
	FF	1*			

Note: An astrick (*) denotes a missed phenotype and or assessment.

This *NEWBORN SCREENING QUALITY ASSURANCE PROGRAM* report is an internal publication distributed to program participants and selected program colleagues. The laboratory quality assurance program is a project cosponsored by the **Centers for Disease Control and Prevention (CDC)** and the **Association of Public Health Laboratories**.

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