MEMORANDUM

TO: Pediatric Providers

FROM: Kathy Cummons, M.S.W.
Office of Maternal, Child and Family Health
Division Director, Research, Evaluation and Planning
Newborn Screening Program

ISSUE DATE: February 21, 2014
RE: Severe Combined Immunodeficiency Disease

EFFECTIVE DATE: March 1, 2014

Effective March 1, 2014, the West Virginia Bureau for Public Health is establishing the expectation that all infants born in a licensed health care facility be screened for severe combined immunodeficiency disease (SCID) or commonly called "Bubble Boy Disease". This screen is in addition to the 30 heritable disorders already screened for in West Virginia as recommended by the U.S. Department of Health and Human Services, Secretary's Advisory Committee on Heritable Disorders and Genetic Diseases. This disorder will be screened for using the newborn blood spots already being collected at the hospital before discharge and submitted to the State Laboratory.

Screening for SCID will also identify other immunodeficiency disorders. This is just a reminder that infants with a positive screen should not receive any rotavirus vaccines until the infant's confirmatory results are obtained. Infants with an unsatisfactory screen should be re-screened as soon as possible.

If any medical provider did not receive newborn screening results for infants under their care, they may call the Office of Maternal, Child and Family Health, Newborn Screening Program at (304) 558-5388, or toll free at 1 (800) 642-8522 or the Office of Laboratory Services/State Lab at (304) 558-3530. Please find enclosed a pamphlet regarding SCID and Rotavirus Vaccines.

KC/vc

Enclosure

cc: Anne Williams
Christina Mullins
Sharon Cibrik
Raheel Khan

OFFICE OF MATERNAL, CHILD AND FAMILY HEALTH
Bureau for Public Health
350 Capitol Street, Room 427
Charleston, WV 25301-3714
Phone: (304) 558-5388 Toll-Free (in WV): 1-800-642-8522 or 1-800-642-9704 FAX: (304) 558-4984
What is SCID?

Severe combined immunodeficiency (SCID) is a genetically-determined condition that results in lack of cellular and humoral immunity. It is estimated that there is one new case per 50,000 to 100,000 live births. Infants with SCID are highly susceptible to infections and may not be diagnosed until these infections prompt the pediatrician to examine their immune system, usually after 4 months of age.

Thus, most SCID infants may have already received one to two doses of the live rotavirus vaccine before the diagnosis is made.

How is SCID diagnosed?

Without newborn screening, the diagnosis of SCID is usually based on history of infections, tests on blood lymphocytes and gene sequence analysis. In only about 20% of cases is a family history of SCID available.
Why and When are Rotavirus Vaccines given?

Rotavirus is a pathogen that usually infects children 6 to 24 months of age. It can cause gastroenteritis with severe diarrhea and dehydration that may lead to hospitalization and even death. Immunization with rotavirus vaccine has been proven to reduce disease severity and mortality from rotavirus gastroenteritis.

The Advisory Committee on Immunization Practices (ACIP) from the Centers for Disease Control and Prevention (CDC) recommends the administration of rotavirus vaccines (2-dose Rotarix® or 3-dose RotaTeq®), starting with the first dose at age 2 months. Rotavirus vaccines are to be given to all infants EXCEPT those with the following conditions:

1) Previous severe allergic reaction to the vaccine or its components.
2) Infants with history of intussusception
3) Infants with ongoing gastroenteritis
4) Severe Combined Immunodeficiency (SCID)

What should a provider do before administering the Rotavirus Vaccine?

1) Review contraindications (e.g. allergies, acute diarrhea) and history of infections.
2) Review family history for suspected immunodeficiency.
3) When available, check the number and percentage of lymphocytes in a CBC, because most SCID patients present with significant lymphopenia (ALC <2,500 cells/mm3).
4) If the CBC indicates lymphopenia, consider withholding the vaccine pending an immunologic investigation.

Providers should follow up all infants receiving rotavirus vaccines for persistent diarrhea and should consult a clinical immunologist if necessary. Even if SCID newborn screening is adopted, some infants with non-lymphocytopenic immunodeficiencies may experience prolonged vaccine-strain rotavirus diarrhea.

Why are Rotavirus Vaccines contraindicated in Severe Combined Immunodeficiency (SCID)?

Rotavirus vaccines are live attenuated rotavirus strains. Infants who are immunocompromised because of SCID may experience severe gastroenteritis caused by rotavirus vaccines, MMR, varicella, and FluMist vaccines are other common live viral vaccines that should not be given to children with severe defects in T cell function.

Reports of five cases of prolonged and severe gastroenteritis caused by rotavirus vaccines in undiagnosed SCID infants were published between September 2009 and January 2010. Four additional cases have been reported to the Vaccine Adverse Event Reporting System (VAERS).

Rotavirus vaccine infection was confirmed in all patients by molecular methods. Based on these reports, the CDC included SCID in the list of contraindications for the administration of the rotavirus vaccines in June 2010.

Recently, SCID newborn screening has been demonstrated to be feasible using dried blood spot Guthrie cards. The U.S. Department of Health and Human Services endorsed the addition of SCID to the uniform newborn screening panel in May 2010. Many states are currently evaluating whether to offer this test, which would keep an otherwise beneficial public health measure, rotavirus immunization, from causing harm to infants with SCID.