Texas Department of State Health Services
1701 N. Congress Avenue, Austin, Texas
Travis Building, T6-128G
October 25, 2013 via Conference Call
Minutes

Conference Call

William Morris, LVN
Alice Gong, MD
Mark Lawson, MD
Elizabeth Stehel, MD
Nancy Beck, MD
Kelly McDonald
Charleta Guillory, MD

Staff

Patrick Clynch, Department of State Health Services (DSHS), Newborn Screening Unit
David R. Martinez, DSHS, Newborn Screening Unit
Daisy Johnson, RN, DSHS
Rachel Lee, DSHS Laboratory, Biochemistry & Genetics Branch
Patty Hunt, DSHS Laboratory, Metabolic Screening
Susan Tanksley, PhD, DSHS, Laboratory Operations Unit Manager

Guests

Ada Drozd, Public Health Department, Texas Medical Association
Laura Blanke, Texas Pediatrics Society
Emily Babcocke, Texas Pediatrics Society
Elizabeth Sjoberg, Texas Hospital Association

Call to Order

Chairman Morris called to order the October 25, 2013 meeting of the Newborn Screening Advisory Committee at approximately 9:00 am.

Roll call of committee members, staff and guests

Chairman Morris asked that everyone take the opportunity to introduce themselves. He reminded everyone that the meeting was being recorded and to identify themselves when speaking. Members, staff and guests attending are listed at the beginning of these minutes. David R. Martinez wanted to take the opportunity to introduce a new member to the Newborn Screening Unit, Karen Hess. He stated that she is the new Newborn Screening Branch Manager, and she is managing the teams for Clinical Care Coordination. He wanted to share some new information with the committee. The unit has been reorganized since expansion. There had been two teams of staff following abnormal screens. There are now three teams, Hemoglobins, Cystic Fibrosis (CF) team, MSMS, Severe Combined Immunodeficiency (SCID) team and an Endocrine team. These three teams make up the branch under Karen's direction. We welcome Karen who managed the Vaccine for Children Program for nine years with DSHS, and prior to that she worked at the Austin/Travis County Health Department managing the Immunization Program and Refugee Program.

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Review and Approval of Minutes

Chairman Morris asked the committee if they had looked at the minutes from the April 26, 2013 meeting and whether any changes needed to be made. Dr. Beck made a motion to approve the minutes. Dr. Stehl seconded the motion. Motion passed.

Newborn Screening Advisory Committee By-Laws-Patrick Clynch

Patrick Clynch presented to the committee a draft of the older by-laws with some small changes based on HB 740. HB 740 changes the composition of the advisory committee. The composition of the categories has been modified. In November, 2009, Commissioner Lakey made the decision that the committee would consist of nine members. HB 740 states that the committee will now consist of the following:

- At least four physicians licensed to practice medicine in this state, including at least two physicians specializing in neonatal-perinatal medicine
- At least two hospital representatives
- At least two persons who have family members affected by a condition for which newborn screening is or may be required under this subchapter; and
- At least two health care providers (persons) who are involved in the delivery of newborn screening services, follow-up or treatment in this state

Based on current committee members, the suggestion is to add another licensed physician and another hospital representative bringing the committee total to 11.

The other small change made was to change the quorum from five to six.

A motion was made by Dr. Gong that the hospital representative should be someone who is an administrator, should be someone that works for the hospital system or even an insurer system and is directly involved in decision making.

Dr. Beck agreed that the rationale for this is the need for someone who is actually involved in policy making within an institution that can advise the committee on things that would be problematic in terms of implementing rules into an actual hospital setting. So it needs to be someone who is in an upper level administration who actually will be able to make those sorts of decisions and would know what goes on in other hospitals at that level.

Susan Tanksley wanted to clarify that the statute has to be followed, so she does not think it would be possible to go to someone who is in Blue Cross Blue Shield or involved with insurance unless they are also directly involved with hospital. We understand the intent of hospital administration, but we also have to follow the statute, and so we can take those things into consideration when looking at the applications. We will depend on the medical associations to try to recommend someone they think would fit well within that role. We don't typically get that sort of applicant.

David R. Martinez also wanted to clarify that internally, the committee's recommendations would be considered when reviewing the applications, but the public solicitation for these members will be for a hospital representative. David also stated that they don't have control over who applies. It is an open

application. Again, the recommendations given by the committee will be taken into consideration when reviewing the applications, but the solicitation will go out as a representative of a hospital.

Chairman Morris clarified that the motion made by Dr. Gong is to make a recommendation to the team that is in charge of reviewing the applications, to consider someone who has a hospital administrative role and use the Hospital Association as a resource for soliciting for someone who would be well suited to the position of hospital representative, giving special consideration to policy making and anything that affects the hospital. Dr. Beck seconded. Motion passed.

Patrick asked if there were any more comments or suggestions for changes in the Newborn Screening Advisory Committee By-Laws. Dr. Gong made a motion to accept the changes to the by-laws. Dr. Beck seconded. Motion passed.

Phenylketonuria (PKU) Monitoring-Rachel Lee

Rachel Lee reported on the PKU Dietary Monitoring Program. It is one of the tests DSHS laboratory provides. The laboratory has been providing this test since PKU screening started back in the mid-1960s, so it has been almost 50 years. The purpose of this testing is to serve patients affected by PKU and their families and basically to help them monitor their diets. Also, to help the physician to make sure the patients are compliant for their diets. Mainly, there are two populations of patients that use this program. One is the infants who are just diagnosed with PKU and still trying to see if the diet is working. The other population is for females that are in the child bearing age. When they are pregnant want to make sure they are not hurting the fetus, so they are trying to monitor the diet. Currently, have about 3,000 specimens per year from about 250 different patients. Usually the patients or family members can either collect the specimens themselves or go to the physician's office and collect there. We have been providing this testing for free for the last 50 years; however, there was a change of policy during the last legislative session. We are now required to recover all the costs for testing. The laboratory began billing (cost of $16.61) for this test on September 1, 2013. We have notified submitters and metabolic specialists to let them know what is happening. Concern was expressed by Dr. Gong that physicians would be charged the $16.61 fee for those patients that are not covered under Medicaid and have no insurance. Susan Tanksley said that the Committee could recommend to DSHS that those patients that are not covered by Medicaid or insurance be covered as charity care. This would be similar to newborn screening where if there is a parent who is unable to pay for newborn screening, they don't have insurance and are not covered by Medicaid or CHIP, the physicians are not charged for that. That is covered partly by Title V and partly through GR.

Dr. Gong made a motion that for those who have no funding that the Department consider writing that off as charity care as opposed to billing the provider. Dr. Stehl seconded. Motion passed.

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Newborn Screening Secondary Targets-Karen Hess

Karen Hess stated that currently the state of Texas screens for the 29 core conditions and hearing screening that are recommended on the newborn screening panel. The law requires that Texas screens for secondary conditions, but only if funds allow and there haven't been funds appropriated to implement the secondary screening conditions. Last November, the Advisory Committee requested that the program follow up with the Commissioner on secondary conditions. In response to that, a draft has been put together, and the options will be presented to the Commissioner on implementing the secondary targets.

- Option 1-include in the current DSHS NBS Panel, the 18 secondary target conditions that are detectable as a consequence of existing laboratory testing
  - Additional Staffing-1 Nurse III position and 1 Public Health and Prevention Specialist III (PHPS) position to support this option
- Option 2-Include 18 secondary target conditions in the DSHS NBS Panel and adjust laboratory testing algorithms
  - Additional Staffing-2 Nurse III positions and 1 PHPS III position to support this option
- Option 3-Include 24 secondary target conditions in the DSHS NBS panel and adjust laboratory tandem mass spectrometry (MS/MS) method to detect six additional secondary target conditions
  - Additional Staffing-2 Nurse III positions, 1 PHPS III position and 1 Chemist IV position to support this option
- Option 4-Make no change to the Texas NBS Panel

Karen Hess stated for clarification that all the options will be presented to Commissioner Lakey with costs consisting primarily of the additional staff.

Dr. Gong stated that if information was going to be presented to Commissioner Lakey with costs, then the committee needs the same information before making a recommendation.

This topic was discussed at great length by the committee with different motions discussed; however, this topic was tabled until the next meeting with no final motion made.

Newborn Screening Rules Update-David R. Martinez

David R. Martinez gave an update on the current rules that are going through the process that the committee looked at earlier this year.

Rules have been through public comments.

- Incorporated many of the comments received
- Incorporated parts of HB 740 into the rules
- Last phase of process; scheduled to be published in Texas Register in early December
  - Become effective 20 days after
  - Should be adopted in mid-December
- Incorporated a new subchapter for Critical Congenital Heart Disease (CCHD) screening
  - Included a definition of CCHD

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- Added the language "physician attending a newborn may delegate the responsibilities to any qualified and properly trained person acting under the physician's supervision relating to providing the confidentiality provisions".
- Added the language "CCHD patients will be able to access the newborn screening benefits".
- Expanded scope of the committee has been added into the rules to include each disorder that is listed on the Recommended Uniform Screening Panel; includes newborn hearing and CCHD
- Added the language about the committee requiring a review of additional screening tests

Mr. Martinez stated that it was a long process for this rule making round, but we are turning around and will be proposing subchapter for screening for CCHD. He directed the committee's attention to Attachment E. Feedback has already been received on some of the things that many expect to see in the rules, and he knows that some may be disappointed, but what has been presented is what was added in statute, and basically the rules are very brief. There is a purpose section, a definition section, an exemption from the CCHD screening, have test procedures and standards and also a reporting of confirmed cases to the department. This is the first draft. It has been reviewed by our attorney to make sure statutorily we are doing what has been outlined in HB 740. There are some expectations that the actual results of the CCHD screen will come to the department; however, the way the legislation was written is very limited in how the department will be involved in. When the legislation was going through the process, the department did not receive any funding for CCHD, so we won't be following these screens like we do the bloodspot screens. It is a point of care test. Again, it is very broad, the purpose is really taken from the statute; we have included definitions for birthing facilities, definition for CCHD, Echocardiogram, a definition for health care practitioners, a neonatal intensive care unit (NICU) definition, definition for pulse oximeter and a screening algorithm. The next section was taken straight out of statute, who is exempt from the screen with the exception of 5 and 6 which we added that the newborn has been previously diagnosed with CCHD to account for some prenatal diagnosis and also 6 to take care of post-natal echocardiogram. The house bill language is specific that the department will develop test procedures and standards. In (a) describe that a birthing facility is required to do the screening test for CCHD. The procedures must be consistent with the screening algorithm and any other protocol currently posted, linked or referenced on our website or any other test procedure designated by the department on its website. What we are looking at proposing on doing is having the algorithms on our website rather than included in the rule because that could change and if it did, we would be required to go through the rule making process which we don't want to have to do. We indicate that the standard will be the pulse oximeter will have to be approved by the FDA for hospital use in newborns, and newborns in the NICU must receive the CCHD screening prior to discharge unless they are exempted from the screen under the exemption section of the rule. The reporting section where there is (a) a physician health care practitioner, health authority, birthing facility or other individual who has the information of a confirmed case for which CCHD is required, shall report a confirmed case to the department. The committee is the first to see this draft of the rules. We have told stakeholders we'd like them to be involved on how we develop a form that we would require how those confirmed cases would get to us and the process for that. We have envisioned that information would be faxed to us. We would have a form on our website that can be utilized for reporting those confirmed cases to us. We would then provide that information to our birth defects section of the department, and we would also keep the information for us, so that we can see how many cases

of CCHD are being reported to the department. Again, the statute is very limited on what the department is going to do. Because we are under a tight time frame the legislation went into effect September 1, 2013, and HB 740 did say we could implement as reasonable after September 1, so we would like to get this on the fast track. We need the rules. We are working on getting the screening algorithm that was produced by the TxPOP project of which Dr. Gong and Dr. Guillory partnered with us in a project. TxPOP developed an algorithm and a tool kit. We hope to be able to put their brochure that was developed, the tool kit, 2 presentations, one for physicians and one for nurses regarding CCHD screening, on our website as soon as possible. Mr. Martinez stated that this was the draft as of now. They will get the committee's feedback and will quickly turnaround and send the draft out to stakeholders so that they can get informal comments back to them. We hope to present the proposed rules at the February DSHS Council meeting and go through the rule making process.

Critical Congenital Heart Disease (CCHD)-Alice Gong

Dr. Gong gave a PowerPoint presentation on CCHD.

- CCHD
  - Ductal dependent systemic circulation
  - Ductal dependent pulmonary circulation
  - Complex critical CHD
  - Physiologic changes may occur after birth
  - Present in extremis with low cardiac output and acidosis, multi-organ failure, hypoxic ischemic brain injury
  - Early detection and timely intervention gives you a much better outcome
  - 50% of CHDs not detected
  - 30% of infant deaths from CCHD occur before diagnosis
- Pulse Oximetry
  - Monitors oxygen saturation
  - Can detect mild hypoxemia without obvious cyanosis
  - Easy to do which is why it is currently the tool of choice
- Pulse Oximetry Screening-Evidence
  - 2 separate large prospective screening of 40,000 newborns in Sweden and nearly 40,000 in Germany
    - Sensitivity 62%, Specificity 99.8%
  - A meta-analysis that was performed after these two studies were published
- Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC)
  - 2010 recommended that CCHD be added to the NBS screening panel
  - 2011 Secretary Sibelius endorsed
- National Efforts
  - Maryland first state
  - New Jersey implemented 2011
  - Other states have gotten together and lots of projects going on
  - Opportunity for other states to learn and not have to re-invent the wheel
- Texas

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  - HB 740 passed and it is named Taryn Kennedy, Nash Sievers & Rex Van De Putte Act
  - Requires all newborns of a birthing facility be screened with exceptions
    - Parental refusal
    - Transfer prior to screening
    - Screening previously completed
    - Discharge before 10 hours and referral made
- Potential Barriers
  - Reporting/Tracking/Quality Improvement (QI)
  - Inadequate resources
  - Resistance from some in the medical community
  - Screener
    - Additional work load
    - Education
  - Equipment
    - Probe, pulse oximeters (2 are FDA approved)
  - Patient/Parent
    - False positives, false negatives
    - Delay in discharge
  - Potential transfer to another center
  - Costs and reimbursement
- CCHD Screening Protocol
  - 7 primary targets
    - Hypoplastic Left Heart Syndrome-most common one
  - Secondary screening targets
    - Not consistently detected
- How to Perform Screening
  - Screen after 24 hours when infant is calm and awake
  - Perform in pre-ductal (right hand) and post-ductal (one foot)
  - If < 90%-positive screen, refer
  - If ≥ 95% in either extremity with ≤ 3% difference, Pass
  - If 90-94% in both or difference > 3%, repeat in 1 hour up to 2 times, then refer
- Texas Pulse Oximetry Project (TxPOP)
  - Joint educational initiative
  - Goal-Develop an appropriate implementation
  - Devised and implemented needs assessment of clinical sites
  - Developed an educational plan
  - 13 facilities in South Texas and Southeast Texas
  - Identified a nurse champion at each facility
- Timeline
  - Project completed; filed report with the state

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Severe Combined Immunodeficiency (SCID) Update-Rachel Lee, Karen Hess

Rachel Lee gave an update on the implementation of SCID. Started SCID screening December, 2012.

- Statistics from December 1, 2012 to August, 2013
  - Presumptive positive has dropped to about .21%
  - 548,025 specimens which equals about 280,000 babies
  - Presumptive positive rate average is about .34%, 1,888 were reported abnormal
  - About 320 screen specimens referred
  - 6 cases of SCID
  - 1 case of SCID variant
  - 107 secondary diagnosis that were sent from specialists

Sunset Review-David R. Martinez

David R. Martinez reported that the Health and Human Services Commission and the 5 agencies under HHSC including DSHS will be undergoing the Sunset Review. The process has already started. Public can contact the Sunset Advisory Commission by December 16, 2013 if they would like to submit any feedback on the department. He wanted to bring to the committee's attention that the Sunset staff requested the committee's contact information, so their information was submitted to the Sunset staff. They will be using that information to send an invitation to provide comments about the agency during the Sunset Review process. The Sunset Self Evaluation Report has been placed on our website at http://www.dshs.state.tx.us/sunset.aspx.

50th Anniversary Celebration Newborn Screening-Bill Morris

Chairman Morris asked that the committee come up with some ideas and/or suggestions about the anniversary celebration.

Public Comments

None

Future Action and Agenda Items

- Discuss options for secondary targets with costs

Adjournment

The next meeting will be held on December 20, 2013. Location will be determined at a later date. There being no further business, the meeting was adjourned at approximately 12:00 p.m.