

The Texas Birth Defects MONITOR



A Semi-Annual Data and Research Update

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Legislative Update

At the conclusion of the Texas Legislature's 81st Regular Session, several bills relevant to birth defects surveillance be-

came law. The following summarizes these bills.

HB806: Insurance Coverage of Prosthetic Devices

Requires health benefit plans to provide coverage for prosthetic devices, orthotic devices, including related professional services and repair and replacement coverage equal to the coverage provided under Medicare. The devices covered would be that determined by the physician or podiatrist to be most appropriate for their patient. Signed by the Governor May 13, 2009 and effective September 1, 2009. (See Limb Reduction Defects, Page 4.)

HB1218: Transferability of Electronic Medical Records between Entities

This bill aims to increase efficiency of the state Medicaid program through the transfer of patient information by means of electronic medical records. HB 1218 requires the Health and Human Services Commission (HHSC) to establish a pilot project in at least one urban area to determine the feasibility, costs, and benefits of exchanging secure electronic health information between HHSC and at least two local or regional health information exchanges. The health information exchanges selected for the pilot must be capable of exchanging electronic health information, including information, at a minimum about medications prescribed, about patients receiving benefits administered by HHSC and providers not owned by a single entity. The pilot will involve exchange of a patient's medication history and could involve the exchange of additional health care information. An evaluation of the benefits of the pilot program and the return on investment will be produced by December 2010. Effective September 1, 2009.

SB1612: Velocardiofacial Syndrome (VCFS)

This bill directs all appropriate HHSC agencies to distribute information about VCFS, a condition caused by a microdeletion of the 22nd chromosome, to parents and appropriate service providers when the agency becomes aware of a child who has at least two of the following conditions:

- o hypotonicity
- o communication delay
- o articulation disorder

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- o resonance disorder
- o nasal regurgitation during feeding as an infant with no history of a cleft palate
- o recurrent ear infections as well as diagnosis of cardiac anomaly, feeding disorder, cleft palate, or submucosal cleft palate; or fine motor or gross motor skills delay.

The legislation was proposed due to concerns that some children are not receiving adequate services due to unfamiliarity with VCFS among intervention services providers. Effective September 1, 2009.

HB1672: Retention of Newborn Screening Blood Spot Cards; Disclosure/Destruction Request Forms

Under State law in Texas, every baby is screened at birth and again about one week later for serious metabolic diseases that are asymptomatic at birth (unless the parents refuse for religious reasons). Once screened and if disorders are identified, serious long term effects and death can be prevented if treatment is initiated quickly. The screening test involves taking a tiny sample of the baby's blood from the heel of the foot to create 'blood spots' on a filter paper card. Each baby's blood spot sample is tested at the Department of State Health Services (DSHS) lab.

Beginning in 2002, DSHS began retaining all used blood spot filter papers for quality assurance and research uses, consistent with state and federal law. Analyses of stored blood spots have the proven potential to advance scientific knowledge of how DNA, proteins, and persistent environmental chemicals relate to health outcomes.

A storage site was located at Texas A&M University in College Station. Beginning with deliveries in mid-2002, all specimens were transferred and securely stored there at room temperature, after one year of storage at DSHS. At the time of specimen transport, personal information such as names of baby and parents and addresses are removed. Each specimen has a unique identification number that allows DSHS staff (but not A&M staff) to electronically link information from the blood spot to each child's birth record at DSHS. Identifying information linking a child to a particular bloodspot is not allowed outside of DSHS without advance consent of the child's parent, managing conservator or legal guardian unless otherwise provided by law. One research purpose of this linkage is

to identify epidemiologic causes of specific disorders at a population-based or group level. Epidemiologic studies using blood spots must be approved by DSHS's Institutional Review Board (IRB), which was established to ensure confidential procedures and protect human subjects in research (which includes these bloodspot specimens).

The types of studies permitted by longer term retention of blood spots include the causes of birth defects, childhood cancer, infectious diseases, autism or mental retardation.

Pilot projects that have evaluated the feasibility of retrieving and analyzing de-identified dried blood spots linked to DSHS disease databases included one on genes related to club foot; another on the effect of blood spot storage on assessing infectious causes of congenital hydrocephalus; and one which explored whether prenatal lead exposure in the mother explains high lead levels in infants.

HB1672 requires DSHS to develop a disclosure/destruction request form that will be distributed by health care providers (or others assisting with the birth) to parents at delivery of a child that, if completed and returned, directs the agency to desist from using that child's blood spots for purposes beyond screening for the disease covered by newborn screening laws, and to destroy the filter card within 60 days after the form is received. Upon adulthood, individuals whose test results have been retained can also request that their cards be discarded.

The bill amends Chapter 33 of the Texas Health and Safety Code, and authorizes specific types of uses of the bloodspot cards after newborn screening is completed, and gives parents/legal guardians/managing conservators the right to instruct DSHS to destroy a bloodspot card after screening Effective September 1, 2009.

HB1795: Enhancement of Newborn Screening Program (Greyson's Law)

This bill directs DSHS to add "secondary target" disorders to the already mandatory newborn screening blood panel, to the extent that funding is available. These secondary targets include disorders of organic acid metabolism, disorders of fatty acid metabolism, disorders of amino acid metabolism and hemoglobinopathies not currently among the disorders for which Texas newborns are screened.

Texas Birth Defects Program: 15 Years

BDES by the Numbers

On March 21, 2009, the Texas Birth Defects Registry celebrated its 15th anniversary! In this issue, we take a look at some key indicators of how the program and its environment have grown since its inception in 1994:

- o Completed cases in the Registry: 157,351
- o Number of staff: 61
- o Staff with 10+ years in BDES: 19
- o Clusters Investigated: 100
- o Data Requests Received: 322 (recorded since 1998)
- o Peer-reviewed publications: 143 (See also Texas Center for Birth Defects Research and Prevention, p x.)

Emergency Response

With their presence in various locations around the state and public health surveillance experience, Birth Defects Epidemiology and Surveillance (BDES) Branch staff are well positioned to contribute to the Texas Department of State Health Services' (DSHS) preparation for and response to various public health emergencies.

Examples of BDES Emergency Assistance Tasks:

- o Collection of medical information on individuals who died as a result of Hurricane Ike.
- o Epidemiologic and administrative functions after the hurricane
- o Reviewing and abstracting information from medical records to help identify new cases of the H1N1 flu and to abstract medical information on those diagnosed individuals.

In 2008 and the first half of 2009, BDES staff hours spent emergency preparedness training and activities totaled approximately 1300 and 1000, respectively.

Costs to the program associated with Emergency Response Training in 2008 were estimated to be nearly \$7000.

Research Center

Texas Center Funding Restored

As reported in the December 2009 issue of the Monitor, as a result of the 2008 competitive grant renewal process, the Texas Center for Birth Defects Research and Prevention was one of three Centers that were not allotted funds to continue this research. We are now happy to report that as additional funds became available to the National Center for Birth Defects and Developmental Disabilities, the Texas Center has again been funded and will continue to fully participate for the next five year cycle.

A planning retreat for Texas collaborators has been planned for August, and priorities and long-term plans will be reported here as they develop. For more information contact Mark Canfield, Ph.D. (mark.canfield@dshs.state.tx.us) or Peter Langlois, Ph.D. (peter.langlois@dshs.state.tx.us), phone 512-458-7232.

Recent Publications

- o Browne, M. L., Rasmussen, S. A., Hoyt, A. T., Waller, D. K., Druschel, C. M., Caton, A. R., et al. (2009). Maternal thyroid disease, thyroid medication use, and selected birth defects in the national birth defects prevention study. Birth Defects Research. Part A, Clinical and Molecular Teratology, 85(7), 621-628.
- o Canfield, M. A., Marengo, L., Ramadhani, T. A., Suarez, L., Brender, J. D., & Scheuerle, A. (2009). The prevalence and predictors of anencephaly and spina bifida in Texas. Paediatric and Perinatal Epidemiology, 23(1), 41-50.
- o Canfield, M. A., Ramadhani, T. A., Shaw, G. M., Carmichael, S. L., Waller, D. K., Mosley, B. S., et al. (2009). Anencephaly and spina bifida among Hispanics: Maternal, sociodemographic, and acculturation factors in the national birth defects prevention study. Birth Defects Research. Part A, Clinical and Molecular Teratology, 85(7), 637-646.

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From the Registry

Focus On: Limb Reduction Defects

Limb reduction defects are relatively common, affecting about 6 pregnancies for every 10,000 live births in Texas. These defects vary quite a bit in severity and can affect both the upper (arms, hands) and lower (legs, feet) limbs. Data from the Texas Birth Defects Registry that comprise upper limb reduction defects include total or partial absence of any portion of the limb. For example, part of the upper arm and hand may be present while the lower arm is absent, as in phocomelia.

Risk factors associated with higher rates of offspring with limb reduction defects include:

- Maternal Conditions: Obesity, pregestational diabetes
- Maternal Exposures: Retinoids, nitric oxide synthase inhibitors (such as the amino acid L-arginine), valproic acid, smoking, thalidomide
- Exposures or Conditions of Both Parents: Exposure to pesticides, consanguinity

- Conditions or Exposures of the Pregnancy: Fetal hypoxia due to mother's use of phenytoin, lacking hemoglobin, amniotic rupture sequence, chorionic villus sampling

Patterns in Texas

Upper Limbs

Relatively few patterns are observed for demographic variables for upper limb reduction defects. Most noticeable is the higher rates among mothers age 40 or older (Figure 1) and among babies born to mothers who live in Dallas-Ft. Worth area (Texas Health Region 3) and lower rates in the Houston area (Texas Health Region 6) (Figure 3).

Lower Limbs

Unlike upper limbs, lower limb defects do not demonstrate a clear association with older maternal age, nor have statistically significant differences emerged for differences in sex of the baby, ethnicity, or border residence. Higher rates are seen in the San Antonio area (Texas Health Region 8) and lower rates in the Houston area (Texas Health Region 6). (Figures 4-6).

Announcements

- Healthy People 2020 Goals Under Development. Every 10 years, the U.S. Department of Health and Human Services leverages scientific insights and lessons learned from the past decade, along with new knowledge of current data, trends, and innovations. Healthy People 2020 will reflect assessments of major risks to health and wellness, changing public health priorities, and emerging issues related to our nation's health preparedness and prevention. Goals and measures established by the Healthy People processes are often used in prioritizing funding for public health activities. Opportunities for public input and to follow the process of developing these important goals can be found at www.healthypeople.gov/.
- Transition Experience of Texas Youth with Disabilities. This monitoring report, published by the Texas Health and Human Services Commission as directed by H.B. 1230, is available at www.hhsc.state.tx.us/reports/HB1230_0509.pdf.
- Weight Gain during Pregnancy: Reexamining the Guidelines has been issued by the Institute of Medicine. It presents specific, updated target ranges for weight gain during pregnancy, including a specific range of recommended gain for obese women. Copies can be pre-ordered at the National Academies Press website, www.nap.edu.
- Project REDD (Research and Education on Disability and Disaster) web site now offers disaster research and recovery resources. Project REDD is housed at the Center on Disability and Development at Texas A&M University. Included are presentations on topics such as disaster planning, the effects of Hurricane Katrina on people with disabilities, and preparing care kits for special education classrooms: <http://redd.tamu.edu>.
- WIC Package Changes: On October 1, 2009, the Texas WIC Nutrition Program food packages will undergo a number of changes. New allowable foods will be added and there will be changes in some of the current food items. These modifications align the WIC food packages with the Dietary Guidelines for Americans and current infant feeding practice guidelines of the American Academy of Pediatrics as well as provide better promotion and support for the establishment of long-term breastfeeding and better accommodation of cultural food preferences. A summary of changes can be found at www.dshs.state.tx.us/wichd/approved_foods/ap_home.shtm.

Reduction Defects of the Upper Limbs in Texas

Reduction Defects of the Lower Limbs in Texas

Figure 1: Birth Prevalence by Maternal Age and Sex of Infant/Fetus

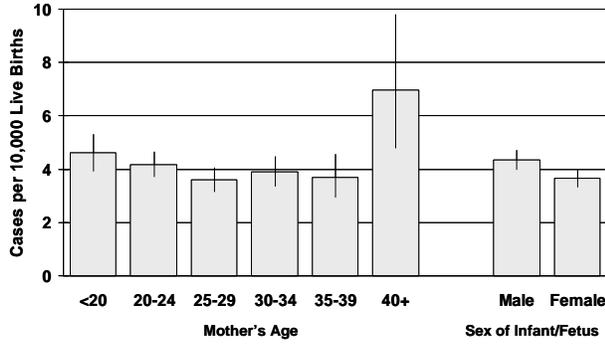


Figure 4: Birth Prevalence by Maternal Age and Sex of Infant/Fetus

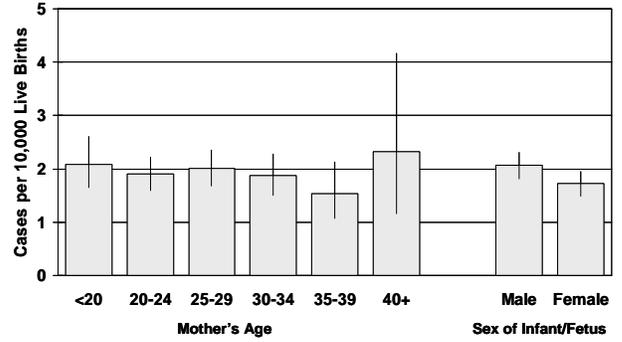


Figure 2: Birth Prevalence by Ethnicity and Border

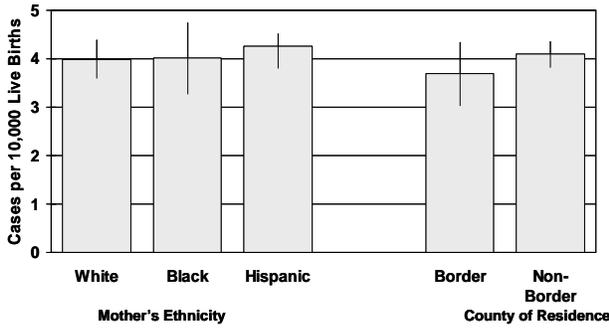


Figure 5: Birth Prevalence by Ethnicity and Border

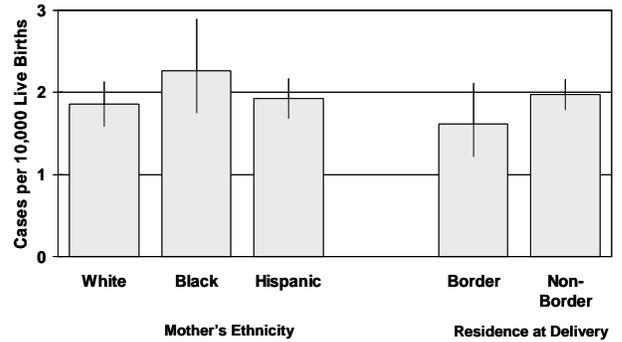


Figure 3: Rates by Texas Health Region

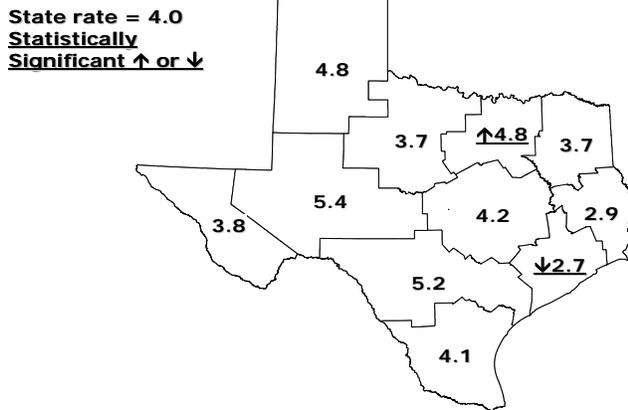
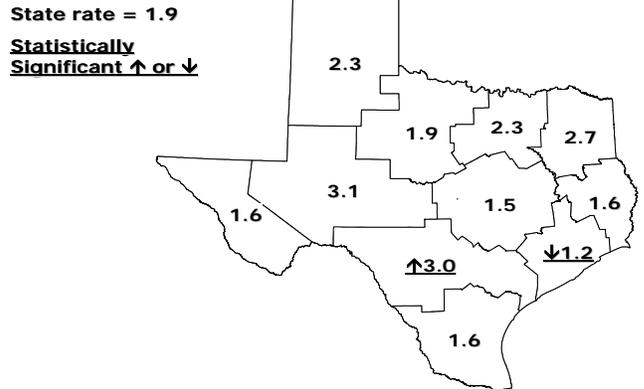


Figure 6: Rates by Texas Health Region



Prevention

NTD Recurrence Prevention Outreach Launched

Neural tube defects (NTDs) occur in approximately 1 per 1000 births in the general population. However, once a woman has had a pregnancy with an NTD, her risk for recurrence in a subsequent pregnancy is 30-50 times higher. Studies have shown that daily preconception consumption of 4.0 mg of folic acid can reduce this recurrence risk by approximately 70%, yet only 33% of women with a previous NTD-affected pregnancy regularly take folic acid.

Previous efforts to prevent recurrent NTDs in Texas concentrated, intense efforts on areas with relatively high rates of NTDs. While these efforts proved effective, they were expensive and did not cover large much of the at-risk population in other parts of the state. In July 2009, however, a new initiative was launched to provide targeted and timely information for women throughout the state at high risk for NTD recurrence. Through this initiative, an educational brochure was mailed to all women in the Texas Birth Defects Registry (about 2500 for the years 1996-2009) informing them of the need for various levels of folic acid supplementation to reduce their NTD recurrence risk. The brochures were designed to reach women at various stages of readiness for adopting this health behavior and employed focus groups of mothers of children with spina bifida to get specific input on appearance, tone, and content.

The Texas Teratogen Information Service is providing phone support for women who receive the brochures and have questions. They are also sending samples with their annual mailing to some 7000 health care providers and pharmacists throughout the state.

In the future, monthly lists of cases of neural tube defects, with mother's most recent mailing address will be generated from completed cases in the Texas Birth Defects Registry and brochures will be sent to them as soon as possible after delivery. For more information, contact Amy Case, 512-458-7232, amy.case@dshs.state.tx.us.

Texas Women's Health Program

The Texas Women's Health Program (WHP) is a Section 1115(a) demonstration waiver enabled by S.B. 747, 79th Legislature, Regular Session, 2005, and approved by the Centers for Medicare and Medicaid Services (CMS) on December 21, 2006. The demonstration started January 1, 2007 and will end December 31,

2011. The Texas Health and Human Services Commission (HHSC) Medicaid/CHIP Division is managing the demonstration.

The WHP is designed to enhance women's health-care services by increasing access to Medicaid family planning for women who have limited health-care resources. Benefits of the program include an annual family planning exam, contraceptives, and related health screenings. The target population is uninsured women ages 18 to 44 with a net family income at or below 185 percent of the federal poverty level (FPL) that would not otherwise be eligible for Medicaid.

Improving access to contraception and providing counseling on the spacing of births through WHP is expected to minimize the overall number of births paid for by Medicaid. For women whose poverty limits their access to health-care services, WHP could reduce the number of infant deaths and premature and low-birth-weight deliveries attributable to closely spaced pregnancies. Improved access may also reduce future disability costs for children arising from premature and low-birth-weight deliveries. For more information on benefits and eligibility requirements for WHP, please visit: www.hhsc.state.tx.us/womenshealth.htm.

Living with Birth Defects

A new report, *Breaking the Link between Special Health Care Needs and Financial Hardship* discusses the prevalence and depth of financial hardship for families of children and youth with special health care needs and explores state-based financing strategies and programs aimed at reducing the financial strain on families. The report, published by the Catalyst Center, provides data on the scope of the problem, illustrates the impact that coverage and financing gaps have on the lives of real families, and describes promising practices and models. Topics include the primary ways a child's or youth's special health care needs may bring financial hardship to families; state-to-state differences in the way programs that serve children, youth, and families are implemented; and the administrative burden on parents when traditional public and private financing sources do not meet their needs. Examples of specific state programs and financing strategies and selected resources are included. The report has been prepared as a resource for policymakers and to support conversations among advocates, families, payers, and Title V programs at both the state and national levels. The report is available at http://hdwg.org/files/resources/Catalyst_Center_Breaking_The_Link.pdf.

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- Zhu, H., Kartiko, S., & Finnell, R. H. (2009). Importance of gene-environment interactions in the etiology of selected birth defects. *Clinical Genetics*, 75 (5), 409-423.

NBDPS Gene Study Funded

Dr. Huiping Zhu at the Center for Environmental and Genetic Medicine, IBT, Texas A&M Health Science Center has received an R21 award from NICHD titled "Genetic Susceptibility of Neural Tube Defects: Diabetes/Obesity Related Candidate Genes." This is the first Texas NIH grant that will use biological samples from the National Birth Defects Prevention Study.

Research Partner Dies

K.C. Donnelly, Ph.D., head and professor of environmental and occupational health at the Texas A&M Health Science Center School of Rural Public Health since 1999, passed away July 1, 2009, from complications related to cancer. With more than 30 years of experience in basic and applied research, Dr. Donnelly was the associate director for the National Institute of Environmental Health Sciences-funded Superfund Basic Research Program at Texas A&M. His research included environmental exposure studies in Azerbaijan; the Czech Republic; Shanxi, China; and numerous U.S. locations, along with animal and human population studies on population exposures and the genotoxicity of complex chemical mixtures. As a collaborator of the Texas Center for Birth Defects research and Prevention, several of his studies looked at the links between environmental toxins and birth defects.



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Calendar

2009

- September 11-15 36th Annual Meeting of the International Clearinghouse for Birth Defects Surveillance and Research, Salt Lake City. www.icbdsr.org/filebank/documents/AnnualMeeting2009_April09.pdf
- September 23-26: National Association of Neonatal Nurses. Austin. Contact: 800-451-3795, or info@nann.org, www.nann.org/about_us/index.html.
- October 3-8: American Health Information Management Annual

Meeting. Grapevine. Contact: info@ahima.org, (312) 233-1100.

- October 14-16: Texas Environmental Health Association Annual Educational Conference. Austin. Contact: 972-461-9644. www.myteha.org/AnnualConference.htm.

2010

- February 7-9: Centering Healthcare Institute's 3rd National Conference On Group Health Care. San Antonio. Contact: 203-271-3632. www.centeringhealthcare.org/pages/features/natl-conference.php.
- March 8-10: National Birth Defects Prevention Network Annual Meeting, Washington, D.C. Contact: Cara Mai 404-398-4918,



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