The Texas Birth Defects

Monitor

A Semi-Annual Data and Research Update

Volume 17, Issue 1 June 2011

82ND TEXAS LEGISLATURE EMPHASIZES HEALTH INFORMATION

INSIDE THIS ISSUE:

Although the 82nd Session of the Texas Legislature faced a full plate of priority items when they convened on January 11, 2011--including balancing a budget projected to be more than $27 billion in the red and the once-a-decade task of re-districting--the body also deliberated and passed four bills that address the collection and use of health information by the state.

Balancing the protection of private health information with transparency and efficiency of government-funded programs to pay for medical care and protect the public health has always been a crucial balance. However, in the past decade, the increasing volume of data and methods for improving its use in research, coupled with a revolution in electronic medical information, has resulted in a need for greater legislative oversight reflected in the following bills adopted this session:

SB156 amends the Texas Health and Safety Code, sec. 108.013 to authorize the Department of State Health Services (DSHS) to share data records with patient identifiers collected from hospital discharge reports (not included in the public use data) with other programs in the agency, allowing for linkage between public health databases. This change has the potential to streamline operations, reduce duplication of effort, and improve research. DSHS programs will be required to have these disclosures reviewed and approved by the department's institutional review board (IRB). Confidential data disclosed to DSHS programs remain subject to all of the applicable policies and statutes. Disclosure of physician-identifying data is not authorized. The bill also authorizes DSHS to share certain confidential data with other health and human services agencies, provided the receiving agency has appropriate confidentiality controls in place. Effective 9/1/1.

SB229 amends Health and Safety Code 47.001, relating to newborn hearing screening in birthing facilities. Under the revised law, both reporting requirements and parental consent procedures are strengthened. Effective 9/1/11.

HB300 creates a Taskforce on Health Information Technology which will develop recommendations to improve informed consent protocols for the electronic exchange of protected health information (PHI); improve patient access to electronic health files for personal health; and address related critical issues. The law also

(Continued on page 6)

FROM THE REGISTRY

10 YEARS OF STATEWIDE DATA

Data are now available on birth defects among deliveries to Texas residents between 1999-2008. The Texas Birth Defects Registry initially began collecting data in a limited geographic area with 1994 deliveries, but did not become consistently statewide until 1999.

The newly published report contains important demographic information for more than 153,000 infants and fetuses affected by one or more major malformation or chromosomal disorders. This represents a nearly unprecedented resource in terms of statistical power, which allows for more meaningful research into the risk factors and causes of individual, and sometimes rather rare, conditions. Furthermore, the scope of these data now allow for studies which include sophisticated investigation of important issues such as whether birth defects rates are changing over time.

The report can be downloaded at www.dshs.state.tx.us/birthdefects/data/BD_Data_99-08/Report-of-Birth-Defects-Among-1999-2008-Deliveries.

Beginning with 2011 deliveries, the Texas Birth Defects Registry added its 200,000th case.

In addition to representing an important resource for research aimed at improving the lives of children, this number emphasizes the scope of the problem of birth defects. We look forward to the day when research and prevention efforts greatly reduce the number of cases added to the Registry each year.

TRISOMY 18 CLUSTER INVESTIGATION

Trisomy 18 (T18) is a chromosomal defect in which an extra copy of chromosome 18 disrupts normal cell functioning; it is usually fatal. Texas Department of State Health Services (DSHS) is investigating a five-infant cluster of T18 conceived during August 2009–June 2010 and delivered during April–December 2010 to residents of Brazos County, Texas. This corresponds to a birth prevalence of 24.2/10,000 live births. In Brazos County and Texas overall during 1999–2007, the prevalence of T18 was 2.4/10,000 live births. This cluster represents a 10-fold increase in T18 prevalence, which was statistically significant ($p < 0.0002$). A 10-fold elevation remained after adjusting for maternal age. One infant was stillborn and three have died.

Community members expressed concern regarding potential health effects from a chemical plant fire that began on July 30, 2009, and burned for 2 days. Tons of ammonium nitrate and possibly other substances were burned. The Texas Commission on Environmental Quality (TCEQ) collected soil and water samples from a location nearby the chemical plant. Metals, nitrates, and organic compounds were tested. Only 4-chlorophenyl phenyl ether and n-Nitrosodimethylamine were higher than their soil protective concentration levels (PCLs). TCEQ's report indicated that they "would not expect adverse health effects to occur" as a result of exposure to these chemicals (unpublished data, TCEQ interoffice memorandum, August 5, 2009).

DSHS is conducting a case-control study of the association between occurrence of T18 and possible risk factors, including exposure to the chemical plant fire. CDC and DSHS are collaborating with Texas A&M University (College Station) researchers to analyze biologic samples from parents of case- and control- infants. Analyses will be guided by results of TCEQ soil and water sampling, published studies of environmental contaminants linked to adverse pregnancy outcomes, and analyses of stored breast milk samples from mothers of two case-infants.

Local physicians reported a perceived increase in spontaneous abortions during 2010. Because chromosomal abnormalities are associated with a higher incidence of spontaneous abortion, Texas A&M researchers are conducting a survey of local physicians to determine the prevalence of spontaneous abortions in Brazos County during 2006–present.

For more information, contact Noha Farag, M.D., Ph.D., EIS Officer, noha.farag@dshs.state.tx.us, (512) 776-7111 ext. 6304.

RESEARCH CENTER NEWS

RECENT PUBLICATIONS FROM TEXAS COLLABORATORS

1. Brender JD, Kelley KE, Werler MM, Langlois PH, Suarez L, Canfield MA: Prevalence and patterns of nitrosatable drug use among U.S. women during early pregnancy. *Birth Defects Res A Clin Mol Teratol* 2011, 91(4):258-264.
2. Browne ML, Hoyt AT, Feldkamp ML, Rasmussen SA, Marshall EG, Druschel CM, Romitti PA: Maternal caffeine intake and risk of selected birth defects in the National Birth Defects Prevention Study. *Birth Defects Res A Clin Mol Teratol* 2011, 91(2):93-101.
3. Case AP, Colpitts LR, Langlois PH, Scheuerle AE: Prenatal diagnosis and cesarean section in a large, population-based birth defects registry. *J Matern Fetal Neonatal Med* 2011.
4. Case AP, Mitchell LE: Prevalence and patterns of choanal atresia and choanal stenosis among pregnancies in Texas, 1999-2004. *Am J Med Genet A* 2011, 155(4):786-791.
5. Davis EM, Peck JD, Thompson D, Wild RA, Langlois P: Maternal diabetes and renal agenesis/dysgenesis. *Birth Defects Res A Clin Mol Teratol* 2010, 88(9):722-727.
6. Duong HT, Shahrukh Hashmi S, Ramadhani T, Canfield MA, Scheuerle A, Kim Waller D: Maternal use of hot tub and major structural birth defects. *Birth Defects Res A Clin Mol Teratol* 2011.
7. Lupo PJ, Langlois PH, Mitchell LE: Epidemiology of Ebstein anomaly: prevalence and patterns in Texas, 1999-2005. *Am J Med Genet A* 2011, 155A(5):1007-1014.
8. Lupo PJ, Symanski E, Waller DK, Chan W, Langlois PH, Canfield MA, Mitchell LE: Maternal exposure to ambient levels of benzene and neural tube defects among offspring: Texas, 1999-2004. *Environ Health Perspect* 2011, 119(3):397-402.
9. Ma C, Carmichael SL, Scheuerle AE, Canfield MA, Shaw GM: Association of microtia with maternal obesity and periconceptional folic acid use. *Am J Med Genet A* 2010, 152A(11):2756-2761.
10. Mei JV, Li L, Rasmussen SA, Collier S, Frias JL, Honein MA, Shaw GM, Lorey F, Meyer R, Chaing S *et al*: Effect of specimen storage conditions on newborn dried blood spots used to assess Toxoplasma gondii immunoglobulin M (IgM). *Clin Chim Acta* 2011, 412(5-6):455-459.
11. Nembhard WN, Salemi JL, Ethen MK, Fixler DE, Canfield MA: Mortality among infants with birth defects: Joint effects of size at birth, gestational age, and maternal race/ethnicity. *Birth Defects Res A Clin Mol Teratol* 2010, 88(9):728-736.
12. Nembhard WN, Salemi JL, Ethen MK, Fixler DE, Dimaggio A, Canfield MA: Racial/Ethnic disparities in risk of early childhood mortality among children with congenital heart defects. *Pediatrics* 2011, 127 (5):e1128-1138.
13. Parker SE, Mai CT, Canfield MA, Rickard R, Wang Y, Meyer RE, Anderson P, Mason CA, Collins JS, Kirby RS *et al*: Updated National Birth Prevalence estimates for selected birth defects in the United States, 2004-2006. *Birth Defects Res A Clin Mol Teratol* 2010, 88 (12):1008-1016.
14. Parks SE, Canfield MA, Ramadhani TA: Importance of including all pregnancy outcomes to reduce bias in epidemiologic studies of neural tube defects--Texas, 1999 to 2005. *Birth Defects Res A Clin Mol Teratol* 2011, 91(3):185-191.
15. Ramadhani TA, Canfield MA, Farag NH, Royle M, Correa A, Waller DK, Scheuerle A: Do foreign- and U.S.-born mothers across racial/ethnic groups have a similar risk profile for selected sociodemographic and periconceptional factors? *Birth Defects Res A Clin Mol Teratol* 2011.
16. Sheu SU, Ethen MK, Scheuerle AE, Langlois PH: Investigation Into an Increase in Plagiocephaly in Texas From 1999 to 2007. *Arch Pediatr Adolesc Med* 2011.
17. Suarez L, Felkner M, Brender JD, Canfield MA: Dieting to Lose Weight and Occurrence of Neural Tube Defects in Offspring of Mexican-American Women. *Matern Child Health J* 2011.
18. Suarez L, Ramadhani T, Felkner M, Canfield MA, Brender JD, Romitti PA, Sun L: Maternal smoking, passive tobacco smoke, and neural tube defects. *Birth Defects Res A Clin Mol Teratol* 2011, 91(1):29-33.
19. Benjamin BG, Ethen MK, Van Hook CL, Myers CA, Canfield MA: Gastroschisis prevalence in Texas 1999-2003. *Birth Defects Res A Clin Mol Teratol* 2010, 88 (3):178-185.
20. Brender JD, Felkner M, Suarez L, Canfield MA, Henry JP: Maternal pesticide exposure and neural tube defects in Mexican Americans. *Ann Epidemiol* 2010, 20(1):16-22.
21. Graham A, Brender JD, Sharkey JR, Zhu L, Felkner M, Suarez L, Canfield MA: Dietary methionine intake and neural tube defects in Mexican-American women. *Birth Defects Res A Clin Mol Teratol* 2010, 88(6):451-457.
22. Griesenbeck JS, Brender JD, Sharkey JR, Steck MD, Huber JC, Jr., Rene AA, McDonald TJ, Romitti PA, Canfield MA, Langlois PH *et al*: Maternal characteristics associated with the dietary intake of nitrates, nitrites, and nitrosamines in women of child-bearing age: a cross-sectional study. *Environ Health* 2010, 9:10.
23. Messer LC, Luben TJ, Mendola P, Carozza SE, Horel SA, Langlois PH: Urban-rural residence and the occurrence of cleft lip and cleft palate in Texas, 1999-2003. *Ann Epidemiol* 2010, 20(1):32-39.

(Continued on page 7)

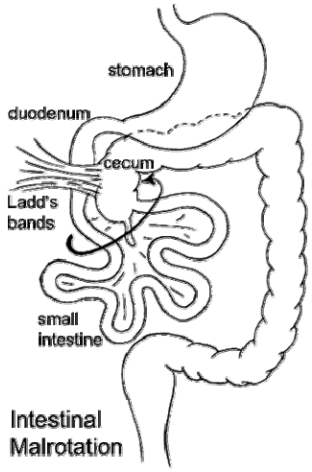
FOCUS ON: INTESTINAL MALROTATION

The primary anatomy and basic structure of the human digestive system are in place by the third month of pregnancy. During the 5th week after conception, the transformation of the embryonic midgut into the mature intestines begins a particularly complex process which includes elongation and positioning of the organ. At this time, the rudimentary intestinal loop emerges from the abdomen through what will become the naval opening, elongates and rotates 90° around its own axis in the process. As this primary intestinal loop grows, so does the liver. Enlargement of the liver pushes the developing intestine into the umbilical cord. During the 7th week the structures return to the abdominal cavity, rotating an additional 180° (total rotation 270° – counterclockwise as viewed from the front). Upon completion, this structure comprises the inferior part of the duodenum, the jejunum, the ileum, the cecum, the appendix, the ascending colon, and the transverse colon. (An excellent description of this process can be found at www.embryology.ch/anglais/sdigestive/mitteldarm01.html.)

Disturbances in this process can lead to several congenital abnormalities, including omphalocele, congenital umbilical hernia, and various defects of intestinal fixation, including intestinal malrotation.

Intestinal malrotation (IM) results from an abnormality of the rotational process: it is actually a failure of the intestine to make or complete the 270° rotation and would more appropriately be called "non-rotation" This non-rotation results in abnormal intestinal placement within the abdominal cavity. Not a medical problem by itself, there can be serious complications because of an intestinal malrotation. Loops of intestine may become bound to other structures, restricting movement and digestion. Malrotated intestine may secondarily twist upon itself (volvulus) resulting in interruption of blood supply and, potentially, necrosis of the intestine itself. These secondary complications may result in severe digestive symptoms and even death in infancy; however, intestinal malrotation may also be asymptomatic or recognized later in childhood or adulthood. Symptoms typically occur if the malrotation process has caused the Ladd's bands (Figure 1) to constrict the duodenum, or if any parts of the intestines become blocked due to twisting upon themselves.

Figure 1



IM frequently occurs in the presence of other birth defects (see below), particularly gastroschisis and omphalocele. (By definition all cases of omphalocele will have a malrotation because of the embryopathogenesis of the defect. For that reason, TBDR does not collect malrotation as a separate defect in the presence of an omphalocele, and we have removed both conditions from the following analysis.)

In the Texas Birth Defects Registry (TBDR), there were 1,153 fetuses or infants with IM (without gastroschisis or omphalocele) identified prior to one year among 1999-2008 deliveries. This represents a birth prevalence rate of 3.31 per 10,000 live births (similar rate to spina bifida). Because IM can have mild or delayed symptoms, this is probably an underestimate of true birth prevalence.

A few statistically significantly variations by demographic group can be observed (Figures 2-3); in particular rates among births to Hispanic women are lower compared to babies born to white and black non-Hispanic mothers (Figure 3).

Nearly 96% of the pregnancies in the TBDR affected by these defects were live born, similar to the proportion of live births for infants with any monitored defect.

There are no national estimates of IM, and the true prevalence in the population is difficult to determine because of the significant possibility of the condition going undiagnosed. However, one study estimated the population prevalence at 1% based on a review of studies examining autopsies of individuals who died from other causes.

Risk Factors

Although virtually no other epidemiologic studies have been published that describe the patterns of this defect in the population, the Hawaii birth defects registry observed similar patterns to Texas, including higher rates among older mothers and

Figure 2: Birth Prevalence of Intestinal Malrotation, Texas, 1999-2008 by Maternal Age and Infant Sex

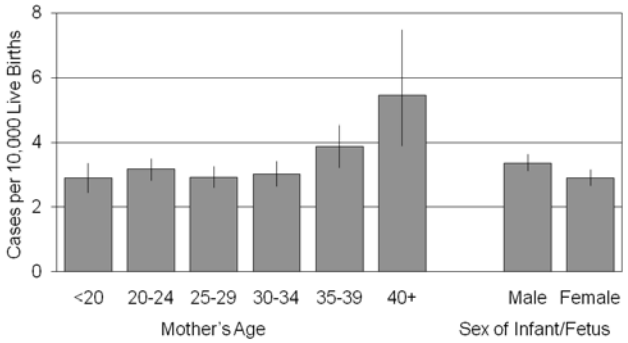
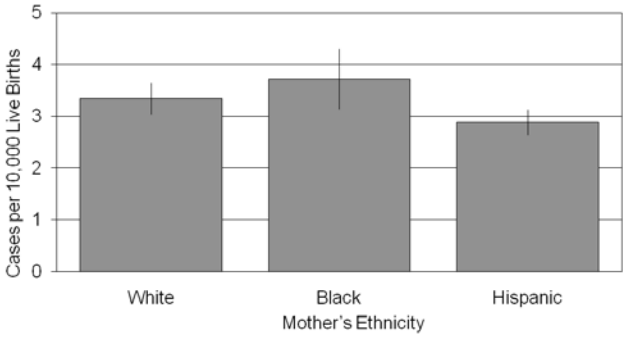


Figure 3: Birth Prevalence of Intestinal Malrotation, Texas, 1999-2008 by Mother's Ethnicity



lower rates among Hispanics. Research remains to be done regarding behaviors, conditions, or exposures that increase the risk for having a child with IM. Some recent research has identified a few genetic variations associated with IM.

Nearly three-quarters of children with IM in the TDBR have at least one additional birth defect. See Figure 4 for other defects that co-occur with IM (in order of frequency).

There are several categories of birth defects which, like IM, can range in severity such that they are immediately recognizable at birth; at the other end of the spectrum, may cause minor or hard-to-diagnose symptoms that are not understood to be due to a birth defect until much later in life*, or not at all.

While data from birth defects systems such as the TBDR cannot identify cases that are diagnosed past infancy, these data provide important clues about the circumstances under which they occur.

Clearly, additional research into the risk factors associated with IM is needed. Additional information about IM can be found at:

http://digestive.niddk.nih.gov/ddiseases/pubs/anatomiccolon/#Malrotation.

**See also, The Cost of Hidden Birth Defects, Texas Birth Defects Monitor Vol. 6, Issue 1, June 2000.*

Figure 4: Defects that Co-occur with IM

Defect	**Number of Cases***
Patent ductus arteriosus	295
Atrial septal defect	239
Obstructive genitourinary defects	211
Stenosis or atresia of the small intestine	206
Ventricular septal defect	181
Atrioventricular septal defect	142
Transposition of the great vessels	126
Pulmonary valve atresia or stenosis	104
Gastroschisis	102
Mitral valve insufficiency	86

*Sums to more than total number of children with IM, because one child may have more than one additional defect.
Italics indicate heart defects.

20 Year Milestone Noted

In April of 1991, a Brownsville, (Cameron County) Texas physician and nurse alerted the Texas Department of Health (TDH) to an apparent cluster of fatal birth defects. The physician had witnessed the birth of three anencephalic infants delivered within a 36 hour period. The reporting of this incident and other similar reports from Brownsville within a six week period sparked what would become a history-making campaign to identify causes of NTDs in Cameron

Impact on Children's Health

Birth Defects Remain Common Cause of Death throughout Childhood

Birth defects are well-known to be the leading cause of infant mortality. However, they remain a significant cause of death among Texans well into young adulthood. In 2008, birth defects were among the ten most common causes of death by age group in Texas for all categories under age 30. The specific ranks and number of deaths attributable to birth defects as a primary cause are shown in the table to the right.

Birth Defects as Cause of Death by Age Group, Texas, 2008

Age Group	Rank	Number of Deaths	% of Deaths in Age Group
<1	1	530	21.4
1-4	2	51	10.1
5-9	3	18	3.8
10-14	5	10	4.0
15-19	6	16	1.5
20-24	7	16	0.9
25-29	9	23	1.3

(Continued from page 1)

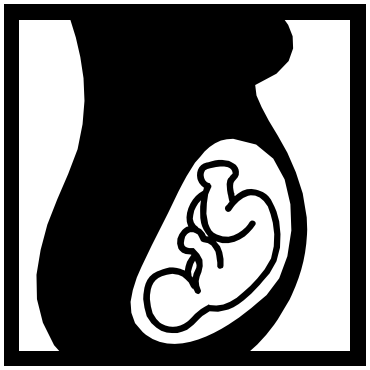
expands the category of "covered entity" when applying federal privacy laws to have the same meaning as it does under Texas statute, which includes public health authorities. Further, the bill prohibits many sales of PHI, and strengthens penalties against individuals and entities violating PHI regulations. Covered entities are now required to notify individuals if protected health information could be disclosed electronically. Effective 9/1/12.

HB411 amends Section 33.0111 of the Health and Safety Code, which describes necessary permissions and procedures for collecting and using genetic material obtained in the screening of all newborns for certain treatable, inherited diseases. HB411 changes this code to only allow public health research using this material, and any information gained from the material with parental express, regardless of whether individual identifiers are excluded in the research analysis. Even with consent for research, the genetic material itself must be destroyed by DSHS two years after receiving it. Effective immediately.

Other legislation of relevance to birth defects includes several laws (below) designed to improve outcomes and accessibility of people with disabilities, including children with birth defects.

HB2610 requires the Health and Human Services Commission to establish a community-based navigator program to assist individuals applying online for public assistance benefits. The program will train and support volunteers and employees of community- and faith-based organizations and will build on similar programs in the Department of State Health Services for training of community health workers. Effective 9/1/11.

HB2636 requires the Texas Health and Human Services Commission to create and appoint members to a Neonatal Intensive Care Unit (NICU) Council. The council will develop NICU operating standards and an accreditation process to receive Medicaid reimbursement. It will also be charged with studying and making recommendations about best practices and protocols to lower NICU admissions. Effective 9/1/11.



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TEXAS CENTER FOR GEOGRAPHIC INFORMATION SCIENCE RECEIVES EPA GRANT

The Texas Center for Geographic Information Science at Texas State University-San Marcos has been awarded $5 million research grant from the US Environmental Protection Agency. The title of the project is "Air Pollution-Exposure-Health Effects Indicators: Mining Massive Geographically Referenced Environmental Health Data to Identify Risk Factors for Birth Defects." The Director of the Center, F. Benjamin Zhan, is the principal investigator of the project. Other co-investigators on the project are Jean D. Brender, Texas A&M Health Science Center, Dr. Peter H. Langlois, Texas Birth Defects Epidemiology and Surveillance Branch, and Dr. Jing Yang, University of North Carolina-Charlotte.

The 3-year project will use data about air pollutants emitted from industrial facilities and birth defects in Texas to develop and evaluate air pollution exposure assessment methods, visual geospatial data mining tools, and epidemiological analysis procedures to define new air pollution-exposure-health effect indicators.

For more information about the project, please contact Dr. Zhan at zhan@txstate.edu.

EDITORIAL NOTE

After more than 13 years as editor and production manager of this publication, I will be leaving my position at the Texas Birth Defects Epidemiology and Surveillance Branch and moving to the Pacific Northwest, where I will be completing my dissertation and providing health communications and policy analysis consulting services.

It has been an honor to be part of such a fine program, and I have appreciated our readers' many comments, constructive criticisms, and compliments. –Amy Case, MAHS

PREVENTION

MOST ANTIBIOTICS HARMLESS DURING PREGNANCY

In June 2011 the American College of Obstetricians and Gynecologists published Committee Opinion #494 "Sulfonamides, Nitrofurantoin, and Risk of Birth Defects," which stated that these antibiotics (commonly prescribed to treat urinary tract infections, or UTIs) are safe for use during pregnancy.

The opinion was issued after researchers from the National Birth Defects Prevention Study (NBDPS) published a report in 2009 indicating increased risk of several birth defects when nitrofurans and sulfomides were taken during the periconceptional period and early pregnancy.

The study measured the association between antibacterial use and selected birth defects. The results of this study indicated that sulfonamides taken between the month prior to conception through the end of the first trimester were associated with anencephaly (adjusted OR=3.4; 95% CI, 1.3-8.8) as well as several other birth defects. Itrofurantoins were associated with hypoplastic left heart syndrome (adjusted OR=4.2; 95% CI, 1.9-9.1), along with 3 other birth defects*.

The Committee cited that since other studies have not observed similar risks, these antibiotics can be prescribed to pregnant women for treatment of UTI's when alternative antibiotics are unavailable.

**Crider, K. S., Cleves, M. A., Reefhuis, J., Berry, R. J., Hobbs, C. A., & Hu, D. J. (2009). Antibacterial medication use during pregnancy and risk of birth defects. Arch.Pediatr.Adolesc.Med, 163, 978-985.*

The Monitor is published twice a year by the Birth Defects Epidemiology and Surveillance Branch, Texas Department of State Health Services:

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Please visit the BDES website for updated information: www.dshs.state.tx.us/birthdefects.

Calendar

2011

- September 24-27: 5th International Conference on Birth Defects and Disabilities in the Developing World. Poland. Contact: icbd2011@firstclass.com.pl
- September 18-24: Deaf Awareness Week. World Federation of the Deaf. Silver Spring, MD. Contact: nadinfo@nad.org
- October 12-15: 14th Annual NPWH Premier Women's Healthcare Conference. Austin, TX. Contact: Carol Wiley cwiley@npwh.org (202) 543-9693
- October 20-21: 12th Annual Chronic Illness and Disability-Transition from Pediatric to Adult-based Care Conference. Houston, TX. Contact: David Madrigal (713) 798-8237; cme@bcm.edu
- November 6-9: 7th Annual International Conference on NTDs. Austin, TX. Contact: vgrier@austin.utexas.edu
- December 14-15: 17th Annual Maternal and Child Health Epidemiology Conference. New Orleans, LA. Contact: cdcinfo@cdc.gov

2012

- January: National Birth Defects Prevention Month. United States. Contact: nbdpn@nbdpn.org
- February 11-14: Association of Maternal and Child Health Programs (AMCHP) 2012 Annual Conference. Washington, DC. Contact: Colleen Campbell ccampbell@conferencemanagers.com (703) 964-1260 ext. 16
- February 27-29: National Birth Defects Prevention Network Annual Meeting, Washington DC. Contact: cwm7@cdc.gov, (409) 498-3918
- April 2-8: National Public Health Week, American Public Health

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