

The Texas Birth Defects  
**MONITOR**



A Semi-Annual Data  
and Research Update

Volume 14, Issue 1

June 2008

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## **Diabetes, Obesity, and Folic Acid Supplementation**

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In Volume 11-1 (June 2005) of this publication, we summarized research pointing to pre-pregnancy diabetes and obesity as risk factors for various birth defects, and have often explored aspects of folic acid and the prevention of birth defects. In 2007, we published a study examining the intersection of these two topics in the Journal

of Obstetric, Gynecological and Neonatal Nursing (Case AP, Ramadhani TA, Canfield MA, Beverly L, Wood R. Folic acid supplementation among diabetic, overweight, or obese women of childbearing age. J Obstet Gynecol Neonatal Nurs. 2007 Jul-Aug;36 (4):335-41). Below is a brief summary of the results.

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While it is known that periconceptional folic acid supplementation can prevent the occurrence of more than 50% neural tube defects (NTDs); and that obesity and diabetes are risk factors for NTDs, there is little known about whether obese, overweight, or diabetic women are equally likely to supplement with folic acid as normal-weight or non-diabetic women. Using data from the Texas Behavioral Risk Factor Surveillance System, we compared responses to the following questions from obese, overweight, or diabetic women to those of normal-weight or non-diabetic women:

- ◆ Do you currently take any vitamin pills or supplements?
  - Are any of these a multivitamin?
  - Do any of the vitamin pills or supplements contain folic acid?
- ◆ How often do you take this vitamin pill or supplement?

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# From the Registry

## Focus On: Achondroplasia

Achondroplasia is a genetic, developmental disorder of the bones and cartilage that results in short stature, lumbar lordosis (sway-back), and limited elbow extension. Commonly called dwarfism, this disorder has several other associated features, including delayed motor development; however, mental retardation is not associated with this disorder. Affected individuals will also have a small foramen magnum (the large hole located in the base of the skull which allows passage of the spinal cord); this can lead to cervicomedullary compression, which may cause infants to have difficulty breathing and sleeping.

This disorder has been found to be autosomal dominant with the vast majority of the cases occurring spontaneously. Children who are homozygous for this disorder are much more severely affected than children who are heterozygous. In fact, the homozygous condition is almost always fatal within a few months after birth.

Affected parents have a 50% chance of having offspring affected with this disorder, and a 25% chance of having a child who is homozygous for this disorder but is physically and functionally unaffected. They also have a 25% chance of having an unaffected child. Recurrence risk for unaffected couples is low.

Advanced paternal age has been linked to this disorder in several studies but has not been linked to advanced maternal age.

Paternal grandparents of children with achondroplasia tend to have higher risk for various types of cancer than do maternal grandparents. This observation has led to a hypothesis on the existence of a "mutator" gene acting in male meiosis and in somatic, mitotic cells in both sexes.

Since achondroplasia is primarily a genetic disorder, there are no known teratogens for this disorder.

In Texas, the rate for achondroplasia is 0.32 per 10,000 live births, but varies somewhat by maternal age and ethnicity (Figures 1 & 2). As would be expected, no regional differences in birth prevalence have been observed in Texas (Figure 3).

Figure 1: Achondroplasia, Texas, 1999-2004

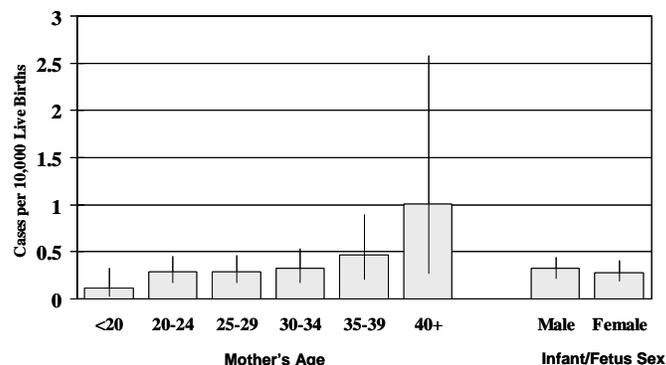


Figure 2: Achondroplasia, Texas, 1999-2004

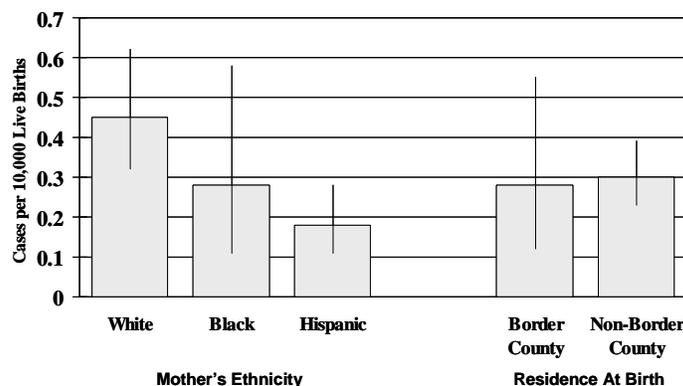
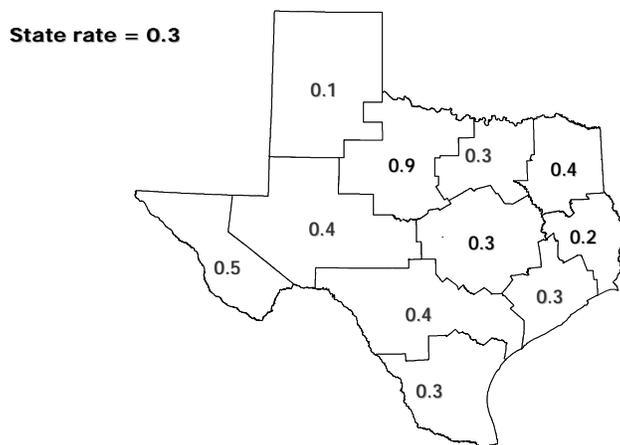


Figure 3: Achondroplasia, Texas, 1999-2004



## Active Surveillance Costs for Birth Defects Registry

In 2007, Birth Defects Epidemiology and Surveillance Branch (BDES) staff visited 228 Texas hospitals and birthing centers to find cases and abstract patient medical records. From these abstractions 20,129 new cases that matched our case definition were found and entered into the Texas Birth Defects Registry. Our data collection staff spent a total of 34,377 hours in travel, research, and data entry for these new cases. The cost of this surveillance in travel dollars and wages was \$625,835. On average, it took surveillance specialists approximately 1.7 hours to find a case and complete an abstraction, at a cost of approximately \$31 for each case. These figures do not include fixed overhead costs such as those associated with program administration and epidemiological analysis.

Compared to 2006, the number of cases abstracted increased by about 10%, but hours per case dropped from 1.9 hours (12%), so total costs for these activities decreased slightly.

BDES staff have developed new tools for measuring and tracking these time and wage costs, which, along with other management indicator data, allow for periodic assessment of program efficiency.

## Prevention

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### Teratogen Fact Sheets Added

The Organization of Teratology Information Services (OTIS) offers many helpful fact sheets regarding potential teratogens as well as non-teratogenic exposures that are often perceived by the public as harmful to the developing fetus. Several fact sheets have been added in the past year: adalimumab (Humira), etanercept (Enbrel), infliximab (Remicade), ciprofloxacin, self tanners, pseudoephedrine/phenylephrine, and research study participation. In addition, fact sheets featuring Bupropion (Wellbutrin) and venlafaxine (Effexor) will be added this summer.

These resources can be found at [http://otispregnancy.org/otis\\_fact\\_sheets.asp](http://otispregnancy.org/otis_fact_sheets.asp) or by calling (866) 626-6847.

## FDA Proposes New Rule on the Use of Prescription Drugs and Biological Products during Pregnancy

The U.S. Food and Drug Administration has proposed major revisions to physician labeling for prescription drugs (including biological products). The proposed changes would give health care professionals more comprehensive information for making prescribing decisions and for counseling women who are pregnant, breastfeeding, or of childbearing age. Under the proposal, drug labeling would explain, based on available information, the potential benefits and risks for the mother and the fetus, and how these risks may change during the course of pregnancy.

Although physician labeling is directed to health care professionals, it is sometimes adapted for use in consumer-directed labeling such as patient package inserts or medication guides when such labeling is approved for a prescription drug.

There are about six million pregnancies in the United States every year, and pregnant women take an average of three to five prescription drugs during pregnancy. Additionally, women with pre-existing medical conditions, such as asthma or high blood pressure, may continue to use prescription drugs to treat those conditions during pregnancy.

In the 1990s, the FDA recognized the shortcomings of pregnancy and breastfeeding information in prescription drug labeling and began reviewing ways to improve the information. The agency held public meetings and focus groups to obtain comments on the current labeling from health care professionals and scientific experts. Current labeling uses a letter category system to describe the risks of drug use during pregnancy, however some stakeholders have said the letter category system leads to an inaccurate and overly simplified view

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of these risks, and does not facilitate updating of labeling as new information becomes available.

The proposed rule would remove the letter categories from the pregnancy section of prescription drug labeling. The newly designed format, for the pregnancy section of the labeling would have three sections:

- ◆ "Fetal Risk Summary" would describe what is known about the effects of the drug on the fetus, and if there is a risk, whether this risk is based on information from animals or humans. The proposal calls for a risk conclusion based on the available data and provides a number of examples depending on the quality and quantity of that data. For example, one risk conclusion might be: "Human data indicate that (name of drug) increases the risk of cardiac abnormalities." This would be followed by a summary of the most important data on the drug's effects.
- ◆ "Clinical Considerations," would include information about the effects of the use of the drug if it is taken before a woman knows she is pregnant. This section also would feature discussions about the risks of the disease to the mother and the baby, dosing information, and tell how to address complications.
- ◆ "Data" would describe in more detail the available data regarding use of the drug in humans and from animal studies that were used to develop the Fetal Risk Summary.

The pregnancy section would also include information about whether there is a pregnancy exposure registry for the drug. Pregnancy exposure registries collect and maintain data on the effects of approved drugs that are prescribed to and used by pregnant women.

Certain newly approved drugs would use the new pregnancy and lactation labeling format, while labeling for previously approved drugs will be phased in gradually under FDA's recent Physician

Labeling Rulemaking. For more information, visit: [http://www.fda.gov/cder/regulatory/pregnancy\\_labeling/default.htm](http://www.fda.gov/cder/regulatory/pregnancy_labeling/default.htm)

## Research Center

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### Recent Publications

- ◆ Brender JD, Suarez L, Langlois PH. Validity of parental work information on the birth certificate. *BMC Public Health* 2008.
- ◆ Brender JD, Zhan FB, Langlois PH, Suarez L, Scheuerle AE. Residential proximity to waste sites and industrial facilities and chromosomal anomalies in offspring. *Int J Hyg Environ Health* 2008; 211(1-2):50-58.
- ◆ Brender JD, Suarez L, Langlois PH. Parental occupation, Hispanic ethnicity, and risk of selected congenital malformations in offspring. *Ethnicity & Disease* 2008; 18(2):218-224.
- ◆ Ethen MK, Ramadhani TA, Scheuerle AE, Canfield MA, Wyszynski DF, Druschel CM et al. Alcohol consumption by women before and during pregnancy. *Matern Child Health J* 2008.
- Husain T, Langlois PH, Sever LE, Gambello MJ. Descriptive epidemiologic features shared by birth defects thought to be related to vascular disruption in Texas, 1996-2002. *Birth Defects Res A Clin Mol Teratol* 2008.
- Yang J, Carmichael SL, Canfield MA, Song J, Shaw GM, National Birth Defects Prevention Study. Socioeconomic status in relation to selected birth defects in a large multicentered US case-control study. *Am J Epidemiol* 2008; 167(2):145-154.
- Felkner M, Suarez L, Liszka B, Brender JD, Canfield M. Neural tube defects, micronutrient deficiencies, and *Helicobacter pylori*: a new hypothesis. *Birth Defects Res A Clin Mol Teratol* 2007; 79(8):617-621.
- Kaye CI, Livingston J, Canfield MA, Mann MY, Lloyd-Puryear MA, Therrell BL Jr. Assuring clinical genetic services for newborns iden-

tified through U.S. newborn screening programs. *Genet Med* 2007; 9(8):518-527.

- Langlois PH, Canfield MA, Suarez L. Are birth defects more prevalent along the Texas-Mexico border? *Texas Medicine* 103[12], 54-59. 2007.
- Langlois PH, Scheuerle A. Using registry data to suggest which birth defects may be more susceptible to artifactual clusters and trends. *Birth Defects Res A Clin Mol Teratol* 2007; 79(11):798-805.
- Mastroiacovo P, Lisi A, Castilla EE, Martinez-Frias ML, Bermejo E, Marengo L et al. Gastroschisis and associated defects: an international study. *Am J Med Genet A* 2007; 143(7):660-671.
- Rasmussen SA, Yazdy MM, Carmichael SL, Jamieson DJ, Canfield MA, Honein MA. Maternal thyroid disease as a risk factor for craniosynostosis. *Obstet Gynecol* 2007; 110(2 Pt 1):369-377.
- Suarez L, Felkner M, Brender JD, Canfield M, Hendricks K. Maternal exposures to cigarette smoke, alcohol, and street drugs and neural tube defect occurrence in offspring. *Matern Child Health J* 2007; 12(3):394-401.
- Wen S, Ethen M, Langlois PH, Mitchell LE. Prevalence of encephalocele in Texas, 1999-2002. *Am J Med Genet A* 2007; 143(18):2150-2155.

## Living with Birth Defects

### CSHCN Waiting List

Effective April 1, 2008, the Children with Special Health Care Needs (CSHCN) Services Program removed 226 clients from its waiting list. As of September 1, 2007, the program has removed a total of 793 clients from the waiting list.

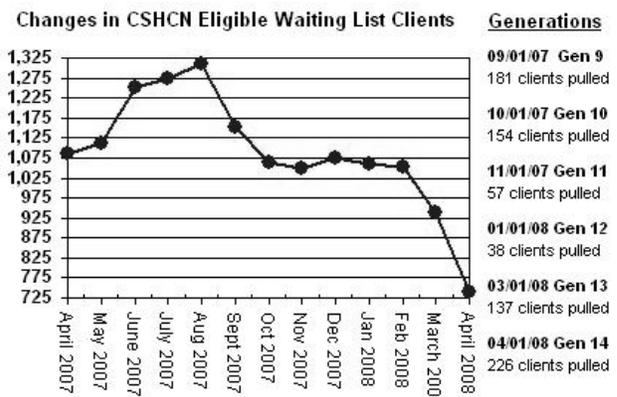
Through state and federal funds, CSHCN provides

health benefits to qualified children with special health care needs and their families, and individuals of all ages with cystic fibrosis. The benefits cover a variety of services, including inpatient rehabilitation physical & occupational therapy, ambulatory surgery, meals, lodging, and transportation needed to obtain medical care, speech and hearing services, medical supplies, and vision care. Medicaid, CHIP, and commercial health insurance benefits, if any, must be used before using CSHCN health benefits.

The CSHCN Services Program places clients on a waiting list for health care benefits whenever there are not enough funds to support all clients seeking health care benefits. The Program may take clients off the waiting list when it has enough funds to provide these clients with health care benefits. The waiting list includes new clients and clients who did not send in a renewal application before their eligibility ran out. All clients including those on the waiting list are eligible for case management services.

There are certain things the Program considers when deciding which waiting list clients may be removed from the waiting list and granted health care benefits, including the doctor's statement on the Physician/Dentist Assessment Form, a client's other insurance coverage, if any, including Medicaid or CHIP, and how long the client has been on the waiting list.

For more information, contact the CSHCN Services Program at 1-800-252-8023.



(Continued from page 1)

- ◆ Some health experts recommend that women take 400 micrograms of the B vitamin folic acid, for which one of the following reasons:
  - To make strong bones
  - To prevent birth defects
  - To prevent high blood pressure
  - Some other reason.
- Has a doctor or nurse ever advised you to take multivitamins or supplements?
- What would you say is the main reason that you do not take any vitamin pills or supplements?

Responses from 6,835 non-pregnant Texas women age 18-44 interviewed from 1999-2003 were analyzed, and crude and adjusted odds ratios were calculated for association between diabetes, body mass index, and folic acid supplementation. While overall 35% reported daily folic acid supplementation we found that obese women were less likely to supplement, even after adjustment for other factors. No other differences were found (Table 1).

All women of childbearing age, but especially those who are obese or diabetic, should be encouraged to take folic acid daily to prevent NTDs. However, no major U.S. professional organization has issued formal recommendations that focus on the special needs for preconception folic acid among overweight, obese or diabetic women. Credentialing and continuing education organizations should be encouraged to incorporate folic acid knowledge objectives into their materials to improve professional understanding of these needs.

**Table 1: Association between diabetes or body mass index and daily folic acid supplementation among women of childbearing age, 1999-2003, Texas**

Characteristics	Crude Odds Ratios	Adjusted Odds Ratios	
		Model 1*	Model 2**
<b>Diabetes</b>			
No	1.00	1.00	1.00
Yes	1.06 (0.77 – 1.46)	1.29 (0.88 – 1.91)	0.95 (0.58 – 1.57)
<b>Sample size</b>	6412	5771	3298
<b>Body Mass Index Category</b>			
Under/normal weight	1.00	1.00	1.00
Overweight	0.91 (0.75 – 1.10)	1.02 (0.79 – 1.32)	0.93 (0.62 – 1.41)
Obese	<b>0.65 (0.54 – 0.78)</b>	<b>0.76 (0.58 – 0.99)</b>	<b>0.65 (0.49 – 0.87)</b>
<b>Sample size</b>	5876	5365	3064
*Adjusted for race/ethnicity, age, education, and household income			
**Adjusted for knowledge that folic acid prevents birth defects and recommendation from a health provider			

# Announcements

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## Genetic Nondiscrimination Information Act Becomes Law

On May 21, 2008 the Genetic Information Nondiscrimination Act (GINA) was signed into law. GINA is the first and only federal legislation that will provide protections against discrimination based on an individual's genetic information in health insurance coverage and employment settings.

The health insurance protections offered by GINA are expected to roll out 12 months after the bill is signed, whereas the employment protections will be fully realized in 18 months.

Specifically, the legislation protects against genetic discrimination by health insurers or employers by:

- Prohibiting group health plans and issuers offering coverage on the group or individual market from basing eligibility determinations or adjusting premiums or contributions on the basis of genetic information. They cannot request, require or purchase the results of genetic tests, or disclose genetic information.
- Prohibiting issuers of Medigap policies from adjusting pricing or conditioning eligibility on the basis of genetic information. They cannot request, require or purchase the results of genetic tests, or disclose genetic information.
- Prohibiting employers from firing, refusing to hire, or otherwise discriminating with respect to compensation, terms, conditions or privileges of employment. Employers may not request, require or purchase genetic information, and may not disclose genetic information. Similar provisions apply to employment agencies and labor organizations.

The Newborn Screening Saves Lives Act also became federal law on April 24. This legislation establishes grant programs, which are to be awarded to eligible entities to provide education in congenital, genetic, and metabolic disorders; and training in newborn screening technologies. Grant programs are also to be used to coordinate follow-up care. In addition to grant programs, this bill increases consumer awareness and knowledge of family support services, research, and other resources

in newborn screening; improves laboratory quality standards; develops a national contingency plan if a public health situation arises; and establishes a central online clearinghouse. Finally, the bill renews the Secretary's Advisory Committee on Heritable Disorders and Genetic Diseases in Newborns and Children, and expands and coordinates research, particularly on conditions that could be added to the panel in the future.

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## Heart Defects: Online Educational Offerings

The National Birth Defects Prevention Network (NBDPN) is now offering free webinars at [www.nbdpn.org](http://www.nbdpn.org) on the following topics:

- The Heart of Surveillance: Tips on Coding
- Syndromes Associated with Congenital Heart Defects,
- Deciphering Diagnostic Tools for Congenital Heart Defects (coming soon)

The presenters are:

- Angela Lin, MD, FAAP, FCMG, Clinical Geneticist, Massachusetts General Hospital for Children, Massachusetts Center for Birth Defects Research and Prevention
- Tiffany Colarusso, MD, FAAP, Pediatrician, Metropolitan Atlanta Congenital Defects Program, Centers for Disease Control and Prevention

These sessions were initially recorded live with a phone-in audience from around the country. Continuing Education Credits are offered for various professions, including physicians, nurses, certified health education specialists, and other interested health professionals. To view the recording and gain access to the handouts, you must be an NBDPN member and log into the Members Only section. (At this time, NBDPN membership is free.)

The NBDPN is a network of birth defects programs and individuals working at the local, state, and national level in birth defects surveillance, research, and prevention.

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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DSHS Pub. No. 58-10955

## Calendar

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### 2008

- ◆ August 8-10: Texas Advocates Conference. Contact: 1-800-252-9729 or (512) 454-6694, [www.texadvocates.org](http://www.texadvocates.org).
- ◆ October: Texas Center for Birth Defects Research and Prevention Symposium, Lubbock. Contact: [amy.case@dshs.state.tx.us](mailto:amy.case@dshs.state.tx.us), 512-458-7232 Ext. 2814.
- ◆ November 3-6: Newborn Screening and Genetic Testing Symposium, San Antonio. [www.aphl.org/profdev/conferences/nbssymp2008/Pages/default.aspx](http://www.aphl.org/profdev/conferences/nbssymp2008/Pages/default.aspx)

### 2009

- ◆ January: Laredo Women's Health Conference. Contact: [events@klrn.org](mailto:events@klrn.org), [www.klrn.org/WomensHealth/LWH08/LWH08\\_Index.html](http://www.klrn.org/WomensHealth/LWH08/LWH08_Index.html)
- ◆ February 23-25: National Birth Defects Prevention Network Annual Meeting, Nashville. Contact: Cara Mai 404-398-4918, [cwm7@cdc.gov](mailto:cwm7@cdc.gov).
- ◆ March 10-12 8th Annual Forum for Improving Children's Healthcare, Grapevine. Contact: 866-787-0832, [www.nichq.org/nichq](http://www.nichq.org/nichq)
- ◆ March 15-18: First World Congress on Spina Bifida Research and Care, Orlando. Contact: Russ Kirby, [rkirby@uab.edu](mailto:rkirby@uab.edu). <http://medicalconference.spinabifidaassociation.org>