An Evaluation of VaCARES: Virginia Congenital Anomalies Reporting and Education System

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The purpose of this report is to disseminate the results of an evaluation of VaCARES. VaCARES is a system in transition, and one of the goals of the evaluation is to inform the grant-funded improvement of the surveillance system and its data collection application, VISITS. Analysis of the system was conducted for current practices and for current data available from VISITS. Recommendations are made in the context of ongoing efforts to revise the system.

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Abbreviations:

DCAH: Division of Child and Adolescent Health
ICD-9-CM: International Classification of Diseases Version 9, Clinical Modification
NBDPN: National Birth Defects Prevention Network
NTDs: Neural Tube Defects
OFHS: Office of Family Health Services
OIM: Office of Information Management at Virginia Department of Health
PPV: Positive Predictive Value
VaCARES: Virginia Congenital Anomalies Reporting and Education System
VaCATPIP: Virginia Congenital Anomalies Tracking and Prevention Improvement Project
VaCHISIP: Virginia Child Health Information Systems Integration Project
VDH: Virginia Department of Health
VISITS: Virginia Infant Screening and Infant Tracking System
VISITS II: Second generation of VISITS, in the process of being rewritten

Public Health Importance of Birth Defect Surveillance

Surveillance, defined as the ongoing, systematic collection, analysis, interpretation, and dissemination of data regarding a health-related event for use in public health action to reduce morbidity and mortality and to improve health (CDC 2001), is an essential service of public health. The surveillance of birth defects allows a state health department to identify risk factors, track local trends, identify clusters, contribute to budget savings, and reduce infant mortality by promoting evidence-based policies and prevention efforts (ASTHO 2004). Ideally, surveillance of birth defects encompasses not only identification of events but also includes prevention activities and follow-up with affected families.

Periodic evaluation of birth defects surveillance systems is necessary to ensure that conditions of public health importance are being monitored efficiently and effectively. The Virginia Congenital Anomalies Reporting Education System (VaCARES) is a statewide population-based surveillance system that collects and reports epidemiologic data on children under age two with birth defects. VaCARES is charged with providing surveillance, education, and follow-up resources for children with birth defects and their families.

Frequency

Although prevalence estimates may vary depending on the number of categories included in the case definition, birth defects are relatively common conditions (Sekhobo and Druschel 2001), with national estimates ranging between 3-5%. March of Dimes reports that the prevalence of birth defects is approximately 3.5% in the United States (MOD 2002). In Virginia, the total number of children born with birth defects for the ten-year period from 1989-1998 was 46,763, with an average of 4,600 children per year. From 1989-1998 the total number of deaths from birth defects, defined as a death from a birth defect before the age of two, in Virginia was 2,724. The prevalence of birth defects in Virginia over the ten-year period from 1989-1998 was 4.9%.

National prevalence estimates vary for specific categories of birth defects. The prevalence of some neural tube defects after fortification of grains with folate, based on data from birth defects surveillance systems with prenatal ascertainment are 3.5 per 10,000 live births affected by anencephaly and 4.1 per 10,000 live births affected by spina bifida from 1999-2001 (MMWR 2004). The national prevalence estimate for Down Syndrome is 14 per 10,000 live births for 1999-2001 based on the birth defects registries of 11 states and adjusted for maternal age (MMWR 2006). The national prevalence estimates for cardiovascular defects are 0.8 per 10,000 live births affected by Truncus arteriosus, 4.7 per 10,000 live births affected by transposition of great arteries, 3.9 per 10,000 live births affected by Tetralogy of Fallot, 4.4 per 10,000 live births affected by Atrioventricular septal defect, and 2.4 per 10,000 live births affected by hypoplastic left heart syndrome for 1999-2001 based on the birth defects registries of 11 states and adjusted for the race-specific distribution of U.S. live births during 1999-2001 (MMWR 2006).

Birth defects have remained the leading cause of infant mortality in the nation for more than two decades (Petrini et al 2002), and are the second leading cause in Virginia in 2002-2004, behind disorders of short gestation and low birthweight. In Virginia, the mortality rate for the ten-year period from 1989-1998 was 29 per 10,000 live births (Li et al 2003).

Though the economic costs of selected birth defects have been studied in detail, there are few estimates of the quality-adjusted life years (QALYs) gained from treatment of all birth defects. Two studies of neonatal surgery, one to treat congenital diaphragmatic hernia (CDH), which is a high mortality condition that requires treatment shortly after birth (Poley et al 2002), and one to treat congenital anorectal malformations (CAM), which is a low mortality condition for which treatment may not result in any gains in continence (Poley et al 2001), assessed quality of life using QALYs. It was found that treated CAM patients gained 13 QALYs over non-treated CAM patients, and that treated CDH patients gained 18 QALYs over non-treated CDH patients. Treated CAM patients have a quality of life (rated by parent as proxy) only slightly lower than the general population (Poley et al 2001), and treated CDH patients had a quality of life rating that was statistically no different than the general population (Poley et al 2002).

Severity

Birth defects may be characterized by severity, as in the scheme used by the National Birth Defects Prevention Network (NBDPN). A major anomaly is a congenital abnormality that requires medical or surgical treatment, has a serious adverse effect on health and development, or has significant cosmetic impact. By contrast, a minor anomaly is one that does not require medical or surgical treatment, does not seriously affect health and development, and does not have significant cosmetic impact. A normal variant is a minor anomaly that occurs in four or more percent of the population (NBDPN 2004).

Among the many types of birth defects there is variation in the severity of morbidity. The most severe can result in death, but many non-lethal defects come with lifelong challenges such as increased hospitalizations and dependence on medical equipment. In Virginia from 1989-1998 on average 260 children died of birth defects every year. The fatality rate for this ten-year period was approximately 5.8%. From 1989-1998, among the 4,676 children born with birth defects in Virginia, 3,839 (82%) required additional hospitalization, and those who needed hospitalization stayed, on average, 11 days (Li et al 2003).

Disparities Associated with Birth Defects

Some disparities associated with the occurrence of birth defects have been noted in Virginia. For the ten-year period from 1989 to 1998, black / African-American children had a higher rate of birth defects than white children (7.1% vs. 4.3%). Children from multiple births were more likely to have birth defects than children from singleton births (9.4% vs. 4.8%). Similarly, children born at low birthweight had more birth defects than children at full birthweight (14.2% vs. 4.2%), children born preterm had more birth defects than children born full-term (10.8% vs. 4.2%), and mothers 35 years of age and older had 3.7 times the risk of having a baby with Down Syndrome than mothers between 20 and 34 years of age (Li et al 2003).

Disparities also exist in the social consequences of birth defects. Physical disabilities may stigmatize a child with birth defects. For example, children with severe spina bifida may have some form of paralysis requiring them to depend on a wheel chair or leg braces for mobility (MOD 2005), may experience difficulty with learning bladder control and rely on catheterization to be able to go to school (SBAA 2005), and may experience social isolation when attempting to integrate with non-affected children.

Costs of Birth Defects

The combined economic cost in 1992 of 18 birth defects (including cerebral palsy), estimated using data from the California Birth Defects Monitoring Program, was eight billion dollars (Waitzman et al 1995). Conditions such as Down syndrome and spina bifida, which are characterized by high levels of long-term activity limitations, had both the highest cost per case and the highest total lifetime costs (Waitzman et al 1995). It is likely that these ten-year old figures substantially underestimate current costs. However, it is clear that cerebral palsy and 17 clinically important birth defects represent an area for considerable cost savings with effective prevention (Waitzman et al 1995).

Birth defects also carry costs other than those considered in traditional economics. The cost of caregiver time and the family impact of birth defects are often not considered when economic estimates are generated. Caring for a child with disabilities can result in changes in economic status for the family, limitations on interaction outside of the family, conflicts within the family, and emotional distress related to the demands of the disability (Tilford et al 2001).

Preventability

Neural tube defects (NTDs) have been the main target of primary prevention efforts in the United States. While some neural tube defects are the result of unalterable genetic factors, the CDC estimates that 50%-70% of NTDs could be prevented by adequate intake of folic acid, at least 400 micrograms of folic acid daily, through dietary intake or supplements (CDC 1992). In September 1992, the U.S. Public Health service issued a recommendation that “All women of childbearing age in the United States who are capable of becoming pregnant should consume 0.4 mg of folic acid per day for the purpose of reducing their risk of having a pregnancy affected with spina bifida or other NTDs” (CDC 1992). In March 1996, the Food and Drug Administration made an announcement requiring that folic acid be added to specific flour, breads and other grains (US FDA 1996). Since that time, numerous follow-up studies have examined the periods pre- and post-fortification to quantify the effects of nationwide fortification and worldwide utilization of supplementation (Bentley et al 1999). Available evidence indicates that fortification has been successful in reducing prevalence of anencephaly and spina bifida, and also some modest reductions in prevalence for some non-NTD birth defects (Canfield et al 2005).

Fetal alcohol spectrum disorders (FASD), which include fetal alcohol syndrome (FAS) and fetal alcohol effects (FAE), can be prevented by not consuming any alcohol while pregnant. The spectrum of disorders ranges from mild disabilities and abnormalities in physical appearance to learning disabilities, growth deficiencies, and nervous system disorders (USDHHS 2005). Similar barriers to prevention of FASD exist as with folic acid because the effects on the fetus happen at the beginning of pregnancy, before a woman may know she is pregnant. The US Surgeon General released an advisory in February 2005, indicating that all women who are pregnant or at risk of becoming pregnant should abstain from drinking alcohol to prevent FASD (USDHHS 2005).

Birth defect surveillance is a necessary step in secondary prevention, both by continuing to uncover the causes of birth defects and also aiding public health personnel in future prevention efforts. Surveillance of birth defects used in conjunction with environmental health data or demographic data can aid in identifying birth defects clusters and targeting interventions and services. Identifying women who had an NTD-affected pregnancy can prevent similar outcomes for future pregnancies through education and nutritional support.

One form of secondary prevention is early detection of birth defects, which can help to minimize morbidity and mortality. Major anomalies, which may be corrected with surgery and physical therapy, require early identification for early intervention. If detected early enough, some defects can be corrected completely so that the child shows no residual effects. Early detection of metabolic syndromes prevents further damage to the body and is crucial to establishing a diet and other therapies that make a long life possible. While surveillance does not directly effect treatment, tracking prevalence of reparable defects helps inform health care providers.

Another form of secondary prevention is prenatal screening for birth defects in the second trimester, which is the standard of care in the United States. First trimester screening for genetic defects has become an option for pregnant women meeting specific criteria. The advantages to first trimester screening include reducing maternal anxiety earlier if the test in negative, allowing women to take advantage of chorionic villus sampling at ten to twelve weeks or amniocentesis in the second-trimester, allowing a woman to prepare for a child with health problems, and affording a woman greater privacy and less health risk in the event that she will terminate the pregnancy (ACOG 2004).

Finally, children with birth defects, their families, and their healthcare providers potentially face a lifetime of treatments that may be difficult to comprehend at the initial diagnosis. Tertiary prevention steps such as necessary treatment, management of conditions, and financial and emotional support are not functions of surveillance systems but are part of comprehensive care for children with birth defects. Surveillance systems can provide follow-up with educational materials and information on support services. In addition, healthcare providers should be able to contact the surveillance system if they suspect a cluster of birth defects or need resources for families.

Public interest

It is difficult to generate general public interest in the prevention of birth defects in part because the public does not want to be reminded that their children could potentially be born with an anomaly, and also because the target population, women who are not yet pregnant, is not necessarily receptive to pregnancy messages, especially if they are not considering pregnancy. A degree of sensitivity is required when handling birth defects prevention. Messages should avoid value judgments about those already living with birth defects. Folic acid campaigns that focus not on birth defects prevention but on promotion of good health have potential for better reaching the general public.

A Birth Defects Awareness Month campaign each January sponsored by NBDPN emphasizes that “It is always a good time to be healthy,” whether or not you are planning a pregnancy. The CDC has slowly shifted health during pregnancy messages to the broader concept of preconception care because many more conditions besides birth defects start with the health of the woman before conception occurs. These messages are slow to come to the public consciousness, but the future of intervention is not at prenatal visits but at pre-pregnancy well-woman exams and clinic visits.

PURPOSE AND OPERATION OF VaCARES

State-Mandated Objectives

VaCARES objectives are mandated in the Code of Virginia (Table 1)

1. To collect data to evaluate the possible causes of birth defects
2. To improve the diagnosis and treatment of birth defects and
3. To establish a mechanism for informing the parents of children identified as having birth defects and their physicians about the health resources available to aid such children.

Planned Uses of the Data from VaCARES

VaCARES has identified several potential uses for surveillance data (VDH 2003). These uses fall into two categories, reporting birth defects data and education about birth defects resources. Reporting is accomplished by identifying birth defect trends among localities and demographic groups, evaluating the success of prevention programs aimed at reducing birth defects in Virginia, and assessing the impact of birth defects on families and communities. Education is accomplished by connecting children with birth defects to resources in their communities, investigating community concerns about birth defects, enhancing public awareness and knowledge, and providing information to scientists that may lead to a better understanding of birth defects. Connecting children with resources requires parent contact for follow-up, which has been suspended because technical issues with VISITS were causing the follow-up letters to be sent later than was useful. Parent contact will be re-initiated when the system regains the ability to identify deceased children in a timely manner to spare families the pain associated with a letter that assumes their child is living. The most recent parent contact letter can be found in Appendix A, p11.

Case Definition

VaCARES uses the following definition for a birth defect: “a serious structural, functional, or biochemical abnormality due to genetic, nutritional, or environmental factors, or any combination of the above, that is present at birth even though it may not be manifested until later in life.” Defects may range from minor to severe and may result in physical or mental disability or death. In this report, “birth defects,” “congenital anomalies,” and “congenital malformations” are used to describe the same conditions (Li et al 2003). To be defined as a case for VaCARES, the following criteria must be met: 1) the child was born alive 2) the child had at least one diagnosis from the VaCARES reportable ICD-9-CM code list reported in the first two years of life and 3) the mother’s residence at the time of delivery was in Virginia. In total there are 86 different categories of structural, functional, or biochemical abnormalities that are required to be reported. The ICD-9-CM diagnosis codes were chosen as the basis for defining birth defects for VaCARES. The ICD-9-CM diagnosis codes for all anomalies meeting the VaCARES definition of a birth defect are listed in Table 2.

Legal Authority

Legislation is required to establish authority to collect data, track cases, and ensure confidentiality, but it can also be a barrier to surveillance if it is not specific enough in defining outcomes or it omits a component of the system (Lynberg et al 1994). The legal authority for data collection and the development of VaCARES was mandated in 1985 and amended in 1986 in the *Code of Virginia*, §32.1-69.1 (Table 1). The legislation creating VaCARES and laying out its mission and objectives is clear and concise.

Organizational Location

VaCARES is housed in the Virginia Department of Health, Office of Family Health Services (OFHS), within the Division of Child and Adolescent Health (DCAH), as part of the Pediatric Screening and Genetic Services program. Virginia Commonwealth University Department of Human Genetics provides expert consultation to VaCARES. The Virginia Genetics Advisory Committee, established under the same law as VaCARES, assists in the ongoing improvement of VaCARES (contact letter Appendix A, p5.). The Virginia Council on Folic Acid (VCFA) is composed of representatives from health and dental professionals across Virginia (contact letter Appendix A, p6). VCFA conducts the Virginia Statewide Folic Acid Campaign in conjunction with the March of Dimes.

Context

There are political, administrative, and social barriers to carrying out birth defects surveillance in any state, and there are some barriers particular to Virginia that must be addressed in this evaluation. Political barriers to surveillance for birth defects include lack of legislation for conducting fetal ascertainment, conservative approaches to family health, and privacy issues surrounding hospital records. Hospital records in Virginia are subject to a strict privacy interpretation by the Attorney General such that one hospital may not see another's records, including those collected by VaCARES. This creates a problem of record duplication, which must be resolved outside of the system by de-duplication and matching with birth certificates.

Administrative barriers to birth defects surveillance exist mainly due to a lack of infrastructure. Contributions of employee time to the system are often "in-kind" donations because there are no full-time employees devoted solely to VaCARES. Birth defects issues cross the lifespan of women, requiring a multi-disciplinary approach to prevention and surveillance across many divisions within family health. For example, the major preventive measure for NTDs is folic acid supplementation for pre-conception and pregnant women, which is handled in the Division of Women and Infants Health, but surveillance of NTDs through VaCARES is handled in the Division of Child and Adolescent Health. While multi-disciplinary collaboration can be beneficial, the lack of permanent staff and funding for surveillance and evaluation efforts leaves the program susceptible to staff turnover, changes in management, and intrusion of other priorities.

Social barriers to birth defects surveillance exist because of the sensitive nature of the topic. One of the program staff refers to this as the "skeleton in the closet". No parent wants to imagine that his or her child will be born less than perfect. VaCARES often uses the term "congenital anomalies" to lessen the stigma associated with the term "defect." The causes of birth defects can be attributed to genetic factors, dietary and environmental factors, or unknown causes. The complex nature of the interaction of these factors and the terminology needed to discuss them further alienates the public. Cultural mythology about pregnancy and births, from predicting the sex of the baby to predicting whether or not it will have a cleft, are barriers to the science of the process, especially where science is not able to provide an alternate explanation.

Renewed privacy issues arose in relation to the collection of protected health information in statewide surveillance systems after the Health Insurance Portability and Accountability Act was enacted in 1996. Even with protection of confidentiality written into the law creating VaCARES, privacy is a core issue for birth defects surveillance. The law supports the surveillance system at the state level, but assurance of parents falls to those who interact with patients.

Level of Integration with Other Systems

The Virginia Infant Screening and Infant Tracking System (VISITS) is a web-based database that integrates surveillance and screening data. Along with VaCARES data, Early Hearing Detection and Intervention (EHDI) data are entered directly into VISITS. Data from birth and death certificates filed with the State Registrar of Vital Records must be loaded into VISITS.

Aspects of prevention and surveillance span various divisions at VDH. The Virginia Council on Folic Acid, within the Division of Women and Infants Health, runs the Statewide Folic Acid Campaign, which distributes materials on folic acid for health care providers and educators and for women of childbearing age. They combine pregnancy-focused ("Healthy Mother Have Healthier Babies with Folic Acid") and healthy women-focused ("Why Every Woman Needs Folic Acid") messages in English and Spanish. In addition, they have co-sponsored, with Divisions of Child and Adolescent Health and WIC and Community Nutrition Services, the distribution of folic acid supplements to family planning and walk-in pregnancy test clinics in 19 Virginia health districts (VDH 2005).

Flow chart of VaCARES

The flow chart documenting VaCARES can be found in Appendix A, p2. The main inputs to the system are when a child is identified through hospital discharge or by the newborn screening system. All data are entered into the VISITS system, and all reports come out of that system. VaCARES is set up as a relational database system with a main data table for the clients, and many tables that house various events for a given client. A flow chart showing how each of these tables is related is found in Appendix A, p3.

Population under surveillance

Birth population. The Commonwealth of Virginia is a mid-Atlantic state encompassing 40,767 square miles, with a large variance in population density and racial composition between urban and rural areas. According to the 2005 Census Estimate, Virginia is the 12th most populous state with 7,567,465 residents. White non-Hispanic residents comprise 69.3% of Virginia's total population and black non-Hispanic residents comprise 20.0% of the total population. While black non-Hispanics comprise less than a quarter of Virginia's population, they are disproportionately represented in infant morbidity and mortality statistics for the state. Females comprise 50.8% of the total population, and reproductive age females (15-44) comprise 21% of the total population.

Birth certificate data for 2004 indicate that there were 103,830 live births to Virginia residents. Selected birth outcomes by race / ethnicity can be found in Table 3. Of the total births for 2004, 60% were to white non-Hispanic females, 21% were to black non-Hispanic females, and 11% were to Hispanic females (any race). In 2004, there were 137,157 pregnancies recorded in Virginia. Of these pregnancies, 25,918 (16%) resulted in induced terminations and 7,409 (4.6%) pregnancies resulted in natural fetal deaths.

Infant mortality. In Virginia from 2002-2004 the infant mortality rate was 7.4 per 1000 live births. There were a total of 2,259 infant deaths, with an average of 753 infant deaths per year. An average of 411 white non-Hispanic infants died annually, resulting in a mortality rate of 5.7 per 1000 live births for 2002-2004. Black non-Hispanic mothers accounted for 22% of all births during

2002- 2004, but 41% of all infant deaths, with an infant mortality rate of 14 per 1000 live births for 2002-2004. In 2004 in Virginia, 67% of all infant deaths occurred during the neonatal period (5 per 1,000 live births), and of 518 neonatal deaths that year, 207 (40%) were black non-Hispanic infants. Of 250 postneonatal deaths in 2004 (2.4 per 1,000 live births), 105 (42%) were black non-Hispanic infants as well, further illustrating the disparity that exists for black non-Hispanic residents of Virginia.

Low birthweight. In 2004, 8.4% of all births in Virginia were less than 2,500 grams and 1.6% of all births were less than 1,500 grams. While the percent of low birthweight for white non-Hispanic and Hispanic infants has remained relatively stable between six and seven percent from 2002-2004, the percent of low birthweight (13%) and very low birthweight (2.8%) black non-Hispanic infants continues to be two times that of infants of other racial/ethnic groups.

Prenatal care. In 2004, 86% of women having a live birth began prenatal care in the first trimester of their pregnancy. Ninety percent of white non-Hispanic women received early prenatal care, while only 79% of black non-Hispanic women entered prenatal care in the first trimester and even fewer Hispanic women (72%) received early prenatal care. Using birth certificate data, the Kotelchuck Index for adequacy of prenatal care utilization indicated that only 80% of pregnant women in 2004 received adequate prenatal care. In 2004 women who had a live birth had an average of 12 prenatal care visits, but 888 women received no prenatal care.

Data Collection

VaCARES data are collected via passive reporting from hospitals on an ongoing basis. Within hospitals, there are designated users who enter eligible discharges via the web-based VISITS system. Data collection was first piloted by Medical College of Virginia in 1986 and then fully implemented at VDH in 1986-1987.

NBDPN recommends collecting some minimum core variables for adequate birth defects surveillance. Table 4 indicates which variables are required to be reported by hospitals when a child with a birth defect is identified in discharge data, which variables are collected from hospitals but are optional, and which variables are supplied from matching to birth certificates.

Reporting Sources for VaCARES

Hospitals are the state-mandated reporting sources for VaCARES. Reporting by university-based genetic centers is optional and is being used to supplement hospital reports. Record duplication is a problem because of the legal restrictions placed on hospital records mentioned in the Context section, but de-duplication the in analysis phase helps to mitigate this problem. In addition to data quality checks in VISITS, the genetic counselors for VaCARES have been doing on- and off-site record reviews to ascertain quality of hospital reporting. These reviews have been used to supplement passive reporting of the hospitals.

Compliance with Standards for Data Formats and Coding

There are no federal standards for birth defects data formatting and coding. In lieu of federal standards, NPDPN has published guidelines pertaining to birth defects surveillance. Compliance with these guidelines is discussed under “Performance of the Surveillance System”.

Data Analysis and Dissemination

Data were analyzed and disseminated in the VaCARES ten-year report by VaCARES and Virginia Commonwealth University staff. Data are analyzed on an ad-hoc basis by various VaCARES staff including the MCH epidemiologist, which will continue until a statistical analyst and a data manager are hired. VaCARES disseminates data yearly to NBDPN.

Policies and Procedures to Ensure Patient Privacy, Data Confidentiality, and System Security

The Commissioner of the Virginia Department of Health maintains the confidentiality of health information collected for VaCARES as described in the *Code of Virginia* §32.1-69.2.

The document “Information Systems: Security and Confidentiality Policies, Procedures, and Standards” serves as the official policies, procedures and standards for Division of Child and Adolescent Health (DCAH) confidentiality and security pertaining to all DCAH data, including VISITS, which contains VaCARES data. The document contains policies regarding physical security of records, disclosure of records, electronic security, and consequences of violations of the confidentiality and disclosure policies. The document is available online and must be received and read by all employees of VDH, contractors, data users, and data recipients who desire access to the VISITS system, which contains person-level birth defects data. De-identified aggregate data may be released by request, and is considered public information. The following statement from the Information Policies document summarizes the data access agreement:

> "DCAH requires signed documentation, utilizing the enclosed Certificate of Receipt for new contractors, data users, and data recipients, and renewed signatures on an annual basis for existing contractors/data recipients, for the purpose of ensuring relevant personnel are initially informed and remain familiar with DCAH policy pertaining to the security and confidentiality of sensitive data."

All those responsible for securing the VaCARES data, including project managers, vendors, the primary information systems developer, and the information systems management contractor, are familiar with the best security practices as described in federal, state, and VDH information systems security laws, policies, standards, directives, regulations, and guidelines and HIPAA proposed IT security regulations which are requirements for the management of secure information (VDH/DCAH 2004). The relevant sections of the *Code of Virginia* and federal regulations regarding reporting and education, confidentiality, and security standards for health information are summarized in Table 5.

Funding sources & Personnel Requirements

The main funding sources for VaCARES are two CDC grants (30%) and Maternal and Child Health Bureau (MCHB) Title V Block Grant funds (70%). The Virginia Child Health Information Systems Integration Project (VaCHISIP) is funded by a CDC grant awarded to improve surveillance at the data level by redesigning VISITS to better integrate the early childhood screening and tracking programs conducted by VDH. The Virginia Congenital Anomalies Tracking and Prevention Improvement Project (VaCATPIP) is funded by a CDC grant awarded to improve surveillance at the system level. MCHB Title V funds mainly support the Genetics Program Manager, the VaCARES Support Technician, and data management and coding.

About five and a half full time equivalent (FTE) staff work with VaCARES. This time is split among approximately 14 people in VDH DCAH and the Office of Information Management (OIM). Salaries are paid through employment at VDH, grants, and contracts with Virginia Commonwealth University.

PERFORMANCE OF THE SURVEILLANCE SYSTEM

NBDPN Guidelines and VaCARES

Virginia recognized the importance of conducting birth defects surveillance, and legislation has facilitated surveillance since 1986. VaCARES has defined objectives and legal authority to conduct surveillance in Virginia. With VaCARES data, VDH is able to describe the prevalence, morbidity, and mortality associated with birth defects. The system is designed to facilitate follow-up of identified birth defect events to provide support and information. The legislation designates VDH as the receiving institution, defines the purpose and objectives of the system, mandates hospital reporting of VaCARES-qualifying discharges, ensures confidentiality and data sharing, and names an advisory committee. The case definition employed by VDH is inclusive of the NBDPN recommendations and goes beyond that to examine other birth defects of importance. The variables collected by VaCARES meet the minimum recommended by NBDPN either through the VISITS system or through linkage with birth certificates. Reporting to VaCARES is based on the five-digit ICD-9-CM coding system. NBDPN recommends using the six-digit CDC coding system, but since hospitals generate discharge reports based on ICD-9-CM, this is the coding system that best facilitates accurate and timely hospital reports in Virginia.

Case ascertainment is passive because VDH receives the reports from the hospitals rather than actively doing chart reviews. While an ideal system utilizes active ascertainment, the passive system employed by VaCARES has proven to be representative of Virginia and hospitals have been making improvements to be compliant with legal mandates. The areas in which VaCARES can improve based on the NBDPN guidelines are 1) data quality (see analysis below), 2) statistical analysis, and 3) data management. Finally, an area where VaCARES can expand current efforts is in dissemination of information. The major effort for dissemination is through NBDPN, which brings state data together to make useful summaries and encourages use of data for planning, prevention, and referral decisions. Overall, VaCARES meets or exceeds the majority of guidelines for conducting birth defects surveillance. The recommendations section at the end of this report explores some strategies for VaCARES to build on the foundation of sound birth defects surveillance.

Level of Usefulness of VaCARES

VaCARES meets most of the applicable criteria indicating the usefulness of surveillance systems set out by the CDC guidelines. VaCARES issued a ten-year report in 2003 that provided estimates of morbidity and mortality related to birth defects, including the identification of associated factors. This analysis and summary can and should be repeated in the future. Although limited by a data collection system that does not lend itself to ad-hoc reporting or timely and regular production of surveillance reports, VaCARES is making every effort to detect trends that signal changes in the occurrence of birth defects, including detection of clusters. In the future the new data collection system will improve data quality and timeliness of surveillance data. VaCARES also permits assessment of the effect of prevention and control programs such as folic acid distribution through WIC clinics for the prevention of Neural Tube Defects. VaCARES staff display a strong commitment to improving the surveillance of birth defects in Virginia in order to improve the quality of life of all children. Improvement of the system will lead to an increase in the use of data to inform policy. The present data collection system has inhibited complex analyses, but the data that are analyzed have stimulated research in the health department, and in the future the data will be accessible enough to facilitate research partnerships with universities. In summary, the new data system will greatly enhance VaCARES staff efforts to collect data to evaluate the possible causes of birth defects, improve the diagnosis and treatment of birth defects, and establish a mechanism for informing the parents of children with birth defects and their physicians about the health available resources.

DESCRIPTION OF SYSTEM ATTRIBUTES

Simplicity

The steps of data flow through VaCARES (Figure A) are: 1) Diagnosis: A doctor makes a diagnosis of a birth defect in a child under two during a hospital stay, a child is diagnosed at a genetic center, or a child is identified through a positive newborn screen. 2) Report: The ICD-9-CM code is identified in a discharge report by the hospital VISITS user and entered into the VISITS system. 3) Analysis: The data in the VISITS / VaCARES system are analyzed at VDH. 4) Dissemination: VaCARES data is reported annually to NBDPN and to Virginia stakeholders at varied time intervals.

Elements of Simplicity. Doctor's diagnosis and / or a positive newborn screen are confirmation that the case definition has been met. The information collected by VISITS / VaCARES is cited in Table 3. Very little maternal exposure data are collected, (alcohol, tobacco, some illicit drugs, agent orange) and this information is not sufficient for in-depth analysis. There is a single receiving organization (VDH). VaCARES is integrated through VISITS with hearing screening, but needs to be better integrated with birth certificates to supply demographic information. All hospitals in Virginia are required by law to report all discharges containing a VaCARES eligible code for a child under two years old. Genetic centers may also report children with birth defects. There is no required follow-up to confirm a "case", but VISITS users are able to contact VDH staff if a record needs to be deleted. As shown in Table 6, most birth defects are confirmed at the time of entry (96%), and there is currently no standard procedure to amend a record. VDH OIM manages the data, and the extent of data management is to ensure that the system is functioning (vs. not functioning). Further data management and cleaning are necessary and will be done when a database manager is hired. The genetic counselor performs quality checks for hospital reporting. Summary data are prepared for annual dissemination to NBDPN. Most recently data were compiled by the MCH Epidemiologist, a DCAH program manager, the CSTE fellow, and the VaCARES Support Technician. Otherwise, data have not been disseminated statewide since the last ten-year report, which was compiled from 1989-1998 data and distributed in 2003. There are no documented methods for data analysis except the guidelines distributed by NBDPN for its annual report. VISITS users undergo training to use the system and to learn the purpose of collecting birth defects for surveillance, and updated training will be initiated for the new system. In the meantime, VISITS hospital users have been involved in user groups that provide feedback on the design of VISITS II. While OIM maintains the current system, VaCARES staff are involved in the design of the new system, which should ensure that VISITS II corrects the difficulties of VISITS, particularly those cited in this evaluation.

The *Code of Virginia* mandates a passive surveillance system, which is simpler than an active surveillance system. However, the simplicity of the surveillance system has been compromised because of the VISITS data system, which has presented a number of difficulties. VISITS was designed through a contract with an outside vendor. Inadequate budget and a host of challenges associated with using an outside vendor (e.g., communication issues, lack of understanding of public health surveillance principles by the vendor) contributed to the lack of efficiency of VISITS. In recognition of these issues, a decision was made to bring the system back to VDH. VDH OIM took over the system and maintains it while a grant-funded rewrite is conducted.

The complexity of VISITS continues to limit the staff’s ability to analyze and disseminate surveillance data. Until the system is re-written, access to data for ad-hoc reporting is limited. Until full time staff (e.g., data manager and statistical analyst) are hired, the data collected will continue to be underutilized. The joint effort between OIM and VaCARES to redesign the data collection system will simplify many of the data issues, and positions are under recruit to foster utilization of the surveillance data.

Data Quality

Methods. *Accessibility of data.* Data were requested from OIM, and available documentation was examined. *Number of birth defects collected*. ICD-9-CM codes for birth defects eligible for reporting to VaCARES (Table 2) were compared with birth defects recommended for collection by NBDPN. *Completeness and validity of entries.* Each of the fields listed in Table 4 that are collected by VaCARES through the VISITS system (indicated by a "+" in the first or second column) were assessed for the number of valid, null (missing), and invalid entries. The fields were categorized as mandatory or optional fields based on information from program staff. Five percent was chosen as a cutoff for non-required fields and required fields that are not essential for analysis.

Results and Discussion. *Accessibility of data.* Data were obtained promptly from OIM once all proper data access and use forms were completed. The data came in Access tables without any documentation. A data dictionary from the vendor that designed the system was available, but the information was incomplete. The documentation did not include the relationships between the tables created in VISITS. In addition, many fields in the tables were not documented in the dictionary, and definitions for field values were often missing.

Number of birth defects collected. A comparison of the VaCARES birth defect diagnosis groups and the major anomalies recommended by NBDPN (NBDPN 2004) revealed that VaCARES included all of the recommended birth defects and more. This is the result of a conscious decision to get a more complete picture of birth defects in Virginia.

Completeness and validity of entries. Table 7 lists fields where fewer than 95% of the entries were complete and valid. Figure B depicts the percent complete and valid for some data fields associated with the child, or client. First name was below the cutoff mainly due to many entries of "BB", "BG", "baby boy", or "baby girl". Users had difficulty distinguishing between plurality (number of infants in a birth event) and birth order (which number child it is), which resulted in making it a non-required field and accounts for the lack of entries. Gender is a required drop-down field, therefore it is complete and valid 100% of the time.

In Figure C, the percent complete and valid for data fields associated with the mother of the client are shown. Mother's name data are of high quality, but mother's maiden name is empty. This is unfortunate because in Virginia, birth records capture mother's maiden name, and this field could have been used to confirm matching, especially where child's first name is missing.

Completeness and validity of entries in contact information fields are presented in Figure D. All fields contain high quality data for this time period. Figures E and F depict the percent complete and valid for data fields associated with hospital information and types of records associated with the client. These analyses aided in identifying which fields had actual information stored and which were empty and no longer useful.

Finally, Figure G depicts the percent complete and valid for data fields associated with race and ethnicity of the client. The labels indicate the variable name in the actual data tables. The lack of documentation for this system resulted in not knowing what definitions were actually associated with the values for race. At first glance, it did not appear that race was being captured by the system. However, race is a required field with a drop-down menu. Through examination of the programming code for reports generated from the "front-end" of VISITS using the available race options, the variables in which race data were stored were determined. Race values are being stored in the field called Ethnicity, and ethnicity, defined as Hispanic or non-Hispanic, is being stored in Ethnicity2, which is not a required field. Table 8 shows the values associated with the original race variable and Table 10 shows the values associated with the original ethnicity variable. The true race and ethnicity definitions are shown in Tables 9 and 11.

Data quality broke down for a number of reasons, including a complex system for data collection, poor to non-existent documentation for the data system, and few error and validity checks. Data quality success occurs for required fields with drop down boxes, such as gender and race, for fields that collect information commonly available, such as current address, for fields that are pre-populated, such as reporting hospital name, and for date fields with a date-picker function, such as date of birth.

Sensitivity

Methods. Sensitivity is the proportion of cases of a disease detected by the surveillance system. In other words, how many children with birth defects are identified by VaCARES out of all who actually have the condition? The years 2000-2002 were chosen for the sensitivity and positive predictive value analyses because these represent three completed birth years of VaCARES data (a completed year for VaCARES data includes data entered up to two years from the date of birth). First, a frequency of births for the years 2000 to 2002 was generated from Virginia resident birth certificates. This frequency was also generated by health district. It is impossible to know how many children in Virginia actually have a birth defect; instead, this number, which is the total of the top row in Table 12, can be estimated by using an assumption of five percent prevalence based on the on the average prevalence calculated in the ten-year report and the known number of live births in each health district. Next, data from birth years 2000-2002 were selected from the VaCARES tables. These data were first de-duplicated by child ID, and then by last name, first name, and date of birth (DOB). This was done to generate a list of VaCARES children with at least one birth defect, since the 5% prevalence estimate is child-based. A frequency of children by health district was generated. The sensitivity calculation can be visualized using Table 12.

Sensitivity = A / (A+C). For this analysis, the estimate of the number of children each health district with a birth defect = A+C, and the frequency of children with birth defects in each health district = A, which is the number of children detected by VaCARES. Sensitivity was graphed for the five health districts that averaged the most live births for the years 2000-2002.

Results and Discussion. The sensitivity of VaCARES for 2000-2002 is increasing over time (Figure H). Figure I depicts the sensitivity trends for the five health districts that averaged the most live births for the years 2000-2002. In four out of the five health districts shown are sensitivity is increasing over time to at or above 100%. In the fifth health district, Virginia Beach, sensitivity has reached only about 50% by 2002.

Sensitivity above 100% does not mean that VaCARES is achieving perfect ascertainment of birth defects cases. There are several explanations for why sensitivity exceeds 100%, and these explanations constitute the limitations of this analysis. The first possibility is that the estimate based on 5% prevalence is an underestimate of the true prevalence of birth defects in Virginia or parts of Virginia. Another reason is that the estimate of expected cases was based only on the number of births in a given health district; the calculation of this estimate did not take into account the race, ethnicity, or socio-economic composition of the health districts, which causes the estimate to be somewhat imprecise.

Finally, there are problems with duplication in the VaCARES database. For this analysis, the records were de-duplicated by child ID (CID) so that a CID with more than one associated defect would not be counted more than once. Once these duplicates were eliminated, the records were then de-duplicated by last name, first name, and DOB grouping. This removes records that have identical names and DOB but different CIDs. However, the records were not de-duplicated to the extent that the same child was entered with a different DOB; even a difference of one number in the date of birth would result in a duplicate record. This problem will be better addressed when the database is formally cleaned and will be minimal once VISITS II is in place and integrated with the birth certificate system. Once duplication problem are minimized, sensitivity can be used to identify areas that have higher numbers of defects than is expected, and cluster analyses of birth defects could then be conducted.

Sensitivity analysis is helpful in identifying low reporting areas, as in Virginia Beach health district. As a result of this analysis, an examination of the reports from hospitals in this health district should be conducted to see if there is a compliance problem, a coding problem, or if there are truly lower numbers of defects in this health district.

Positive Predictive Value

Methods. Positive Predictive Value is the proportion of reported cases that do have the health-related event under surveillance. In other words, how many children have the specific birth defect out of all who were identified by VaCARES? There are several limitations to conducting this type of analysis using a chronic disease. There are very few laboratory tests for confirmation of birth defects, and those that exist are not tracked. VaCARES is based on the hospital discharge record, so using hospital records to confirm birth defects would actually just be using the same source. This is a good check to make sure information is entered properly, but it is not confirmation of a diagnosis. The best independent source would be to have a second physician confirm each birth defect. Since most of the time this is not possible, and it is not tracked when it happens, the next best “confirmatory” source is the birth certificate.

Birth certificates capture birth defects somewhat simplistically with check boxes, but VaCARES is able to capture a higher degree of complexity using ICD-9-CM codes. A crosswalk table was employed to facilitate coding of VaCARES defects the same way as birth certificates (see Table 13). Since birth certificates were the limiting factor, those categories used in the analysis. It is expected that PPV will be higher for defects easily identified at birth (and therefore more likely to be marked on the birth certificate) and for defects that match up with a few specific ICD-9-CM codes.

VaCARES child records were extracted from the VaCARES database with the following data fields: child ID, last name, first name, and DOB. Infant birth certificates with a congenital anomaly recorded were extracted with the following data fields: Unique ID, last name, first name, and DOB. The VaCARES and birth certificate records were matched on last name, first name, and DOB, and a new table was made with both the CID and the Unique ID. The records were then linked back to the VaCARES database and the birth certificate database, and the defect codes associated with the IDs were pulled into the new table. A new variable called “Match” was created and was equal to one when the VaCARES code was equal to the birth certificate code. The PPV calculation can be visualized using Table 12.

PPV = A / (A+B). For this analysis, the total number of VaCARES records for which a matching birth certificate could be found for each defect = A+B, which is a sample of the children caught by VaCARES. The number of records in which the VaCARES code matched the birth certificate code = A, which is the number of children detected by VaCARES and confirmed by birth certificates.

Results and Discussion. Positive predictive values for categories of neural tube defects are shown in Figure J. The categories shown are those listed on the birth certificate. As expected, PPV is higher for Spina bifida and meningocele which are easily identifiable at birth, and hydrocephalus and microcephalus, which both match up to just one ICD-9-CM code each. By comparison, the “other CNS anomalies” category matches up to 10 ICD-9-CM codes and has a much lower PPV. The positive predictive value is shown in Figure K for three categories of what could be called structural birth defects, and one chromosomal anomaly, Down Syndrome. PPV is relatively high here, ranging from about 57% to 72%.

The number of anomalies that would be reported based on birth certificates and VaCARES over four years are shown in Figure L. It is important to remember that although both these sources are considered passive, birth certificates represent only a snapshot of birth defects in Virginia, whereas the hospital discharge reports over two years of a child’s life make for a much more dynamic and informative system. Figure M shows the prevalence resulting from the two sources of birth defects reports. Based solely on birth certificates, the prevalence of birth defects in Virginia would just approach one percent, which does not capture the true situation.

In the context of birth defects, a low PPV highlights how comprehensive the hospital-based system is over birth certificates alone. In the future, a better PPV analysis can be done because VaCARES is recruiting university-based genetic centers whose reports will provide independent confirmatory diagnoses.

It is notable that in Figure N, which shows the numbers of NTDs detected by both sources, anencephalus was recorded more often on birth certificates than in VaCARES. One possible explanation for this discrepancy is that since anencephalus is such a severe defect, the child may not have lived long enough to be admitted to the hospital, and therefore would not have a discharge record. VaCARES collects more defects than recommended by NBDPN, which forces their prevalence closer to the 5% end of the 3-5% range for birth defects. However, collecting more defects than are recommended allows the health department to understand more about birth defects in Virginia. A surveillance system based on hospital discharge reports captures much more information on many more defects than birth certificates alone.

Representativeness

Methods. A mapping approach was taken to determine whether VaCARES was approximately representative of birth defects in Virginia. Records for children with birth defects reported in birth years 2000-2002 were identified and addresses were cleaned for content and completeness. These addresses were then geocoded by VDH Division of Disease Prevention. Upon receipt of the coordinates, the data were loaded into ArcView GIS and maps were generated for all children with at least one birth defect and all children with a central nervous system defect. These maps were overlaid or compared side-by-side with a map of the population of zero to two-year-old children in Virginia from 2000-2002 based on population projections.

Results and Discussion. The top map in Figure O shows the concentration of zero to two-year-old children from 2000-2002, with the darkest areas representing the most densely populated areas in Virginia. The bottom map is a spot map of all cases of birth defects reported in Virginia between 2000 and 2002. There are three areas of high population concentration. They are, from top to bottom, the area around Washington DC including Alexandria, Arlington, and Fairfax, the Richmond area, including Henrico and Chesterfield, and the Norfolk, Portsmouth, Virginia Beach area.

The areas of highest concentration of zero to two-year-old are also where the largest clusters of birth defects reports are located. The map in Figure P shows cases of central nervous system defects, as defined by the NBDPN annual report, for 2000-2002. As before, the clusters of cases reported are in areas of highest concentration of the zero to two-year-old population.

By mapping the birth defects cases, it is possible to visualize whether the surveillance system is approximately representative of the population in Virginia. It will now be possible to identify clusters of defect reports, examine reporting by geography over time, and to look at reports for specific types of defects.

Stability

Methods. Discussions with VaCARES staff about the funding sources of the system, the support for the system, and issues of staffing were used to establish the stability of the system.

Results and Discussion. Funding sources for VaCARES are from CDC grants (30%) and Maternal and Child Health Bureau Title V Block Grant funds (70%). The CDC grants are for a limited period, for the improvement of surveillance at the system level (VaCATPIP: Virginia Congenital Anomalies Tracking and Prevention Improvement Project) and the improvement of surveillance at the data level by redesigning VISITS to better integrate the early childhood screening and tracking programs (VaCHISIP: Virginia Child Health Information Systems Integration Project). When these grant cycles are complete, additional funds will be sought to continue enhancement and data quality. Maintenance of the system “as-is” would be possible without additional funds as long as Title V funds are available. Ideally, additional grants would be secured to move the system towards an active-passive hybrid. It is not likely the CDC grants will be withdrawn because they are non-competitive, so as long as VaCARES improvement continues and all necessary information is provided to the CDC, the grant should be stable for the full term. The costs of the system are absolutely justified and “what we do with the money we have is very good.” More funds would allow VaCARES to do additional prevention activities and to move forward. Converting the system to a hybrid system, or to go completely active requires partnerships with universities and millions in additional funding. The ultimate goal is for VaCARES to become part of the CDC Centers for Birth Defects Research and Prevention Study.

Acceptability

Methods. VaCARES User Survey. VaCARES staff conducted a user survey in February of 2005 to obtain feedback on VISITS in preparation for the design of VISITS II by OIM. The survey was designed to asses users' knowledge of various aspects of the system as well as their opinions on the ease of use of the system. The survey was sent to 303 VISITS users; of these, 124 were Early Health Detection and Intervention Program (EHDI) users, 165 were VaCARES users, 11 were both EHDI and VaCARES users, and three were pilot users for At Risk, a new module in VISITS.

Hospital Reporting. The genetic counselor tracks hospital reporting to detect low reporting rates and to target hospitals for record reviews. A six-year trend report was produced from the Microsoft Access data tables rather than pulling the reports out of VISITS (which, since it is client-based, simply generates a listing output of all children reported by the hospital rather than a summary). This trend report was analyzed and the percent of hospitals increasing their reports from 2000-2003 was calculated.

Results. *VaCARES User Survey: Characteristics of Respondents*. The response rate among VaCARES users was 18%. The majority of VaCARES users categorized themselves as daily, weekly, or monthly users (67%), followed by infrequent users and those who had not yet used the system (33%).

Response to Multiple Choice. The majority of all users agreed or strongly agreed that training was useful (86%) but were uncertain that the user manual for VISITS was helpful (50%). The majority of users seldom or never experienced error messages (78%) or experienced any difficulty connecting to VISITS (77%). The majority of users strongly agreed or agreed that VISITS was easy to understand (82%) and easy to operate (83%). The majority of users had never experienced restrictions when accessing a child's record (79%) and the majority were able to contact an administrator in a timely fashion if they were having problems with

VISITS (65%). *Free Response Questions.* In response to an open-ended question regarding what aspects of the data system were liked the least, some common themes were that the password had to be changed every 30 days, dissatisfaction with the date-picker/calendar, physician selection, and that the system was slow, time-consuming, or had too many screens. In response to the question "Are there any issues at the hospital that prevent you from reporting through the VISITS system in timely and efficient way?" the most common issue was staffing needs and time to enter the data. Suggestions to improve the VISITS system included changing the calendar and eliminating the provider search. Other comments received were positive, with the exception of a complaint about the time it takes to enter cases even when all the information is ready to be entered, and that it was difficult to connect to the VISITS application.

Hospital Reporting. From 2000-2003, 60% of hospitals showed an increase in reporting cases. In that same time period, 16% showed a decrease in reporting, 14% stayed the same, and 10% were considered not eligible for trends because of an OB closure or not yet opened in 2000.

Discussion. Overall, the users who responded were satisfied with the VISITS application. Problems such as the time it takes to enter cases and the ease of connection and use will be addressed in VISITS II. Strengths of the VISITS system are that it is web-based and there is support by VDH for those who have problems. Users did not like the calendar, or "date-picker," which is one of the tools used to make dates higher quality.

Hospital reporting rate can be used as a measure of acceptability because it indicates willingness to comply with state law. The majority of hospitals have increased their reporting from 2000-2003, and this is a trend that is expected to continue over time, especially as the VaCARES staff further utilizes enforcement strategies. The strategies used now are on-site and off-site record reviews by the genetic counselors and annual reports to hospitals with their own progress as well as that of all Virginia hospitals.

Timeliness

Methods. Three aspects of timeliness were examined: 1) hospital reporting time, 2) period of data collection, and 3) dissemination. As part of the genetic counselor's compilation of the VaCARES Annual Report to Hospitals (examples in Appendix D, p1-2), the average reporting time for 2005 reports to VaCARES was calculated. This is defined as the time between discharge of a reportable case and the actual report to VaCARES via VISITS. The expectation of VaCARES staff, which has been communicated through VISITS trainings, is that the infant or child will be reported via VISITS no later than 30 days after discharge. The average reporting time was compared across hospitals. The percent of hospitals in compliance with the 30-day reporting expectation was calculated. To examine whether the lateness of reporting is just a matter of days or more a matter of months, the percent of hospitals reporting within 40 days and over 100 days was calculated.

Results and Discussion. When examining the first aspect of timeliness, hospital reporting, 44% of hospitals in birth year 2005 were compliant with the 30 days reporting expectation. This takes into account only eligible hospitals, which were defined as those who made a report to VaCARES in 2005 (62 hospitals out of 74 non-psychiatric, acute care hospitals). The percent of hospitals reporting within 40 days was 54%, a gain of only 11% over those reporting at 30 days. Finally, the percent of hospitals with a reporting time greater then 100 days was about 13%. Reporting time over 100 days may indicate that hospitals have entered old VaCARES eligible discharges, particularly after a record review. While this indicates these hospitals are attempting to come into compliance with the law, it is still a marker of missed cases and a barrier to timeliness.

A second aspect of timeliness has to do with the period of data collection. VaCARES collects surveillance data on children in Virginia with birth defects from birth until two years of age, which allows the system to catch children with anomalies that are not recognized at birth because the infant is too small, the diagnosis cannot be made at birth, or the specific defect does not fully develop until later. The two-year window helps prevent underreporting of birth defects, but this feature is also a limitation for data analysis and reporting of findings because there is at least a two-year lag for all “final” analyses.

The third aspect of timeliness is dissemination of results. A survey of health district nurse managers highlighted a concern that the ten-year report (1989-1998), which was widely distributed to Virginia’s health districts in 2003, was too outdated to be useful to them (Appendix A, p7). VaCARES has reported birth defects data annually to the NBDPN since 1998, and the reports have appeared in Congenital Malformations Surveillance Reports published in *Birth Defects Research* (Table 14). A strategy to address the concerns of nurse managers is to notify them when the journal article comes out, and to send them program and progress updates on the system redesign. Maintaining partnerships in the field will encourage the use of data when it is released.

Flexibility

According to VaCARES personnel, past changes to the VISITS system, as a result of discovery of errors or a decision that data should be captured in another way, were associated with a fee and could take up to one year to be completed. Based on this information, the data system could be characterized as inflexible. However, the most profound change to VaCARES will be the implementation of the new data system, VISITS II. As a result of extensive planning, careful consideration, and procurement of appropriate funds, it appears the transition to VISITS II will be smooth.

CONCLUSIONS

There are three main challenges facing VaCARES. The first is difficulty with data sharing. A state law prohibited hospitals from seeing other hospitals’ patient records, so the integration of birth certificate data and birth defects hospital discharges was previously precluded. As a result, duplication of records in the system makes analysis difficult. The second challenge is the complexity of the VISITS system. The design of the system and the subsequent transfer of the system from the outside agent to the health department have created data quality issues. Access to data is a major barrier to timely surveillance. The redesign of the system by VDH OIM will alleviate many of the access problems. The third challenge is a lack of full time data management and statistical staff. VaCARES is in need of a data manager and a statistical analyst to facilitate cleaning and use of the available data. These staff are necessary to submit the required reports but also to use the data for surveillance, including geographic analyses, linkage to environmental health databases, and coordination of birth defects research with university partners.

VaCARES and its staff are moving forward to accomplish the stated objectives of surveillance, education, and follow-up. The staff took preliminary results of this evaluation and began making progress almost immediately. Through cooperation with stakeholders, the law has been changed to allow hospitals to see other hospitals' records. Data quality issues have been incorporated into the requirements document for VISITS II. All involved with VaCARES are committed to running a high quality surveillance system and are striving to better serve Virginia children with birth defects and their families.

RECOMMENDATIONS

- **Ensure that the data are used for surveillance as well as follow-up**.
- **Use funds from vacant positions to contract out initial data cleaning activities** until the statistical analyst and database manager currently under recruit are hired.
- **Document the current system**, and ensure that the new system has excellent documentation.
- **Clean the existing data for reporting and research purposes.** The data from VISITS needs to be reported until completed years of data can be reported from the new system.
- **Disseminate birth defect surveillance summaries** (similar to NBDPN report) with counts and rates for most recent completed years of data.
- **Update nurse managers and other key stakeholders** about the redesign of VISITS, what programs are supported by VaCARES, and when new reports will be available.
- **Update website to be both data and consumer friendly**. The Virginia Cancer Registry, another public health surveillance system, has a website

with good layout and content. Items marked with a * can be modeled after the VCR website.

  - Objectives mandated in Code of Virginia
  - Frequently Asked Questions (FAQs)*
  - Basic information like that of the fact sheet (Appendix A, 17-18), but on the front page instead of as a PDF.
  - Assurance of confidentiality of VaCARES data*
  - Link to the most recent data (surveillance update)
  - What the data is used for (and what it is <u>not</u> used for)*
  - Contact for more information, for example: What to do when your child is diagnosed
  - Link to all the fact sheets (examples in English and Spanish in Appendix A, p19-22).
  - Links to Virginia Genetics Advisory Committee, Virginia Council on Folic Acid, and March of Dimes
  - Information for those interested in research using the VaCARES data
  - Consider gathering some “success” stories – how families are coping, what they thought were the best resources

- **Re-initiate parent contact.** This activity addresses a major portion of the objectives mandated by law. Plans to re-initiate parent contact are already under way. The new parent contact letter is in Appendix A, p11.
- **Pay attention to the data entry logic**. Whenever possible, the system should have forced-choice options for data entry to limit errors, provide an “unknown” option, and pre-populate fields, especially when the new system is integrated with the birth certificate system.
- **Make ongoing efforts to evaluate data quality** to catch problems early on, especially when implementing the new system.
- **Geocode and map all reported cases** to supplement current reporting capacity and facilitate cluster investigation.

- **Create a manual for data analysis and reporting birth defects statistics** to encourage researchers to use the raw data and ensure that birth defects statistics are reported accurately.
- **Conduct another surveillance evaluation after implementation of the new system** using the Surveillance Methodology Evaluation Plan (Appendix A, p12-16).

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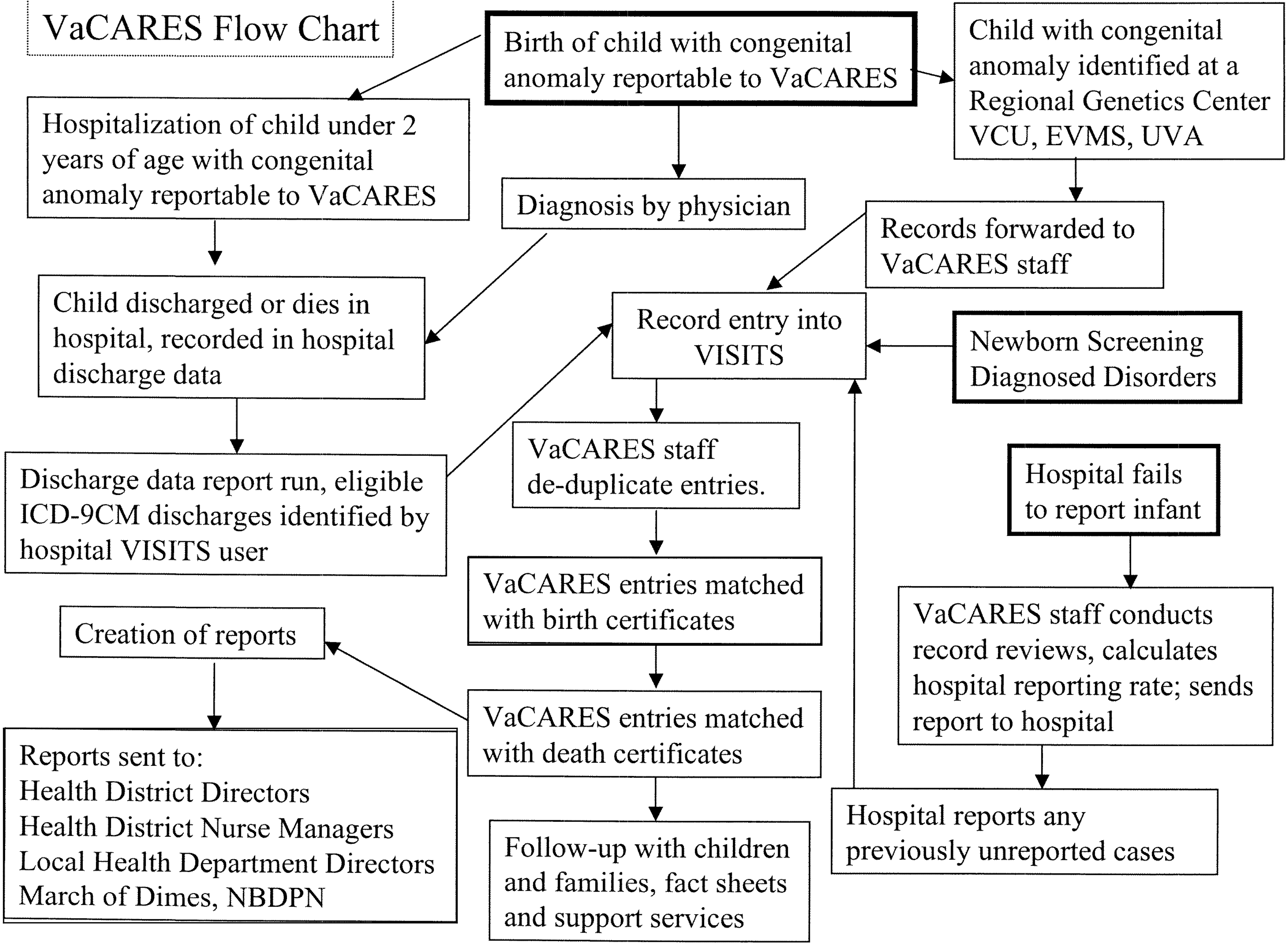
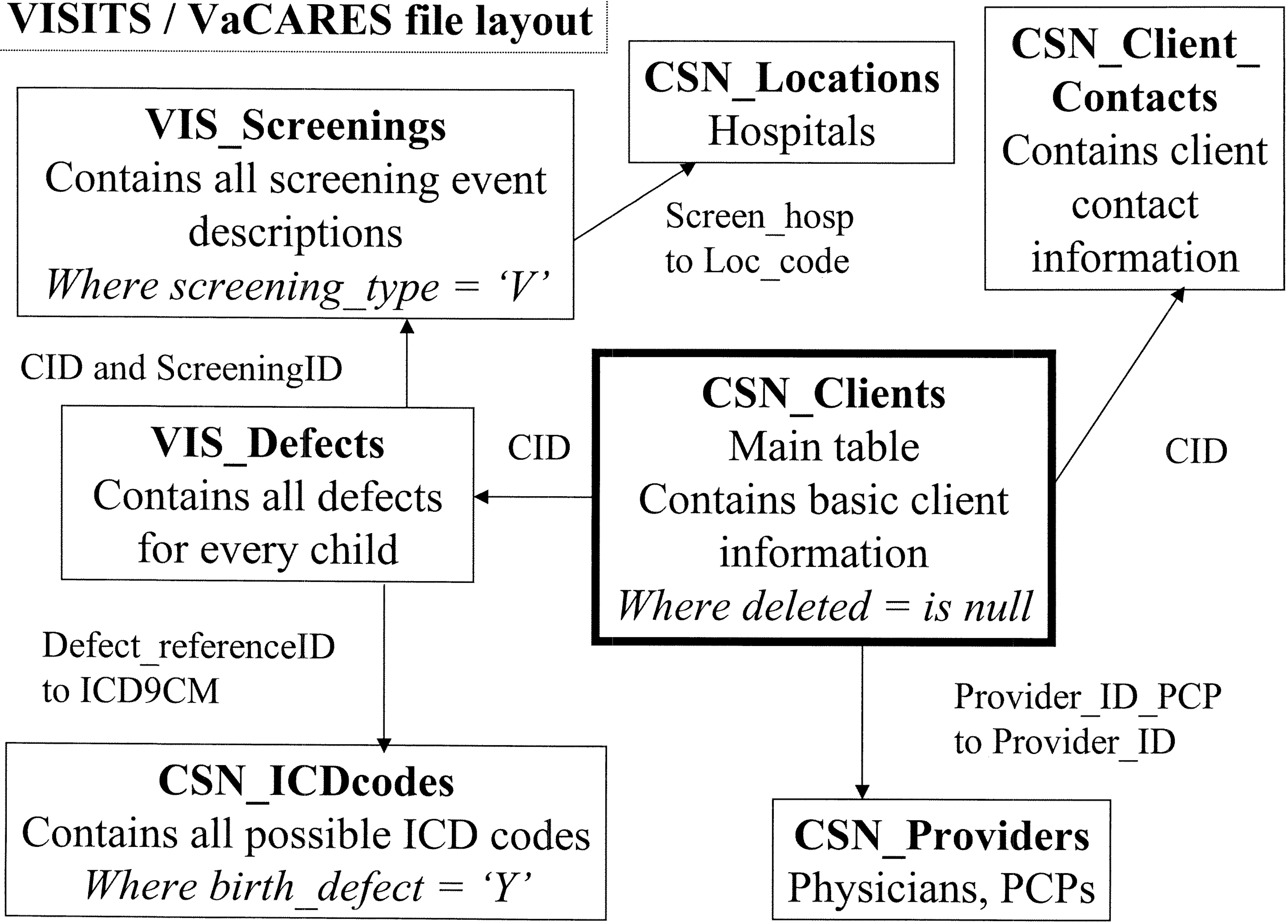
Appendix A –Documents

VaCARES Flow Chart



VISITS / VaCARES file layout



VIS_Screenings
Contains all screening event descriptions
Where screening_type = 'V'

Screen_hosp
to Loc_code

CSN_Locations
Hospitals

CSN_Client_ Contacts
Contains client contact information

CID and ScreeningID

VIS_Defects
Contains all defects for every child

CID

CSN_Clients
Main table
Contains basic client information
Where deleted = is null

CID

Defect_referenceID
to ICD9CM

Provider_ID_PCP
to Provider_ID

CSN_ICDcodes
Contains all possible ICD codes
Where birth_defect = 'Y'

CSN_Providers
Physicians, PCPs

Office of Family Health Services
Date: November 17, 2005

Dear Nurse Managers:

On behalf of the Virginia Department of Health Office of Family Health Services, I would like to inform you of a project I am leading. I am working at VDH through a fellowship sponsored by the Council of State and Territorial Epidemiologists (CSTE) and the Centers for Disease Control and Prevention (CDC). I am conducting an evaluation of Virginia's birth defects surveillance system (VaCARES, the Virginia Congenital Anomalies Reporting and Education System).

Periodic evaluation of surveillance systems is necessary to ensure that problems of public health importance are being monitored efficiently and effectively. Using this evaluation, I plan to make recommendations to those who administer VaCARES for improving the quality, efficiency, and usefulness of the system.

My first task for this evaluation is to identify the stakeholders, those who use the surveillance data for the promotion of healthy lifestyles and the prevention and control of birth defects. As a district nurse manager, you have received the VaCARES 10-year report, 1989-1998. I would like your input on how VaCARES is working for you now.

Would you please fill out the attached survey and email it back to me at your earliest convenience? Upon completion of this evaluation, I will send you the findings so you can see how your contribution has helped VaCARES continue to improve its surveillance and education services. I look forward to working with you.

Thank you for your assistance.

Sincerely,

Caroline C. Stampfel
CSTE Fellow

Office of Family Health Services
December 12, 2005

Dear Members of the Virginia Genetics Advisory Committee:

On behalf of the Virginia Department of Health Office of Family Health Services, I would like to inform you of a project I am leading. I am working at VDH through a fellowship sponsored by the Council of State and Territorial Epidemiologists (CSTE) and the Centers for Disease Control and Prevention (CDC). I am conducting an evaluation of Virginia's birth defects surveillance system, VaCARES.

As a member of the Virginia Genetics Advisory Committee, you have assisted in the design and operation of VaCARES. Periodic evaluation of surveillance systems such as VaCARES is necessary to ensure that problems of public health importance are being monitored efficiently and effectively. Using this evaluation, I plan to make recommendations for improving the quality, efficiency, and utility of the system.

The three main objectives for this evaluation are:

1) To gather information about the functioning of VaCARES with regard to fulfilling the three objectives mandated by state law
2) To evaluate VaCARES using the National Birth Defects Prevention Network's (NBDPN) Guidelines for Conducting Birth Defects Surveillance and the CDC's Updated Guidelines for Evaluating Public Health Surveillance Systems, and
3) To inform the design of a VaCARES Surveillance Methodology Evaluation Plan which will be a protocol for future evaluations.

I have attached a two-page response form to this email to help focus any feedback you may have. I hope you will feel free to let me know of any specific concerns or questions you want to have addressed by the evaluation. Upon completion of this evaluation, I will communicate the findings to you so you can see how your contribution has helped VaCARES continue to improve its surveillance and education services. I look forward to working with you.

Thank you for your assistance.

Sincerely,
Caroline C. Stampfel
CSTE Fellow

Office of Family Health Services
December 14, 2005

Dear Members of the Virginia Council on Folic Acid:

On behalf of the Virginia Department of Health Office of Family Health Services, I would like to inform you of a project I am leading. I am working at VDH through a fellowship sponsored by the Council of State and Territorial Epidemiologists (CSTE) and the Centers for Disease Control and Prevention (CDC). I am conducting an evaluation of Virginia's birth defects surveillance system, VaCARES.

As a member of the Virginia Council on Folic Acid, your role in prevention of neural tube defects (NTDs) through the Virginia Statewide Folic Acid Campaign is an essential companion to VaCARES. The existence of a birth defects registry gives you the opportunity to determine where to target your efforts and how well your program is working. Periodic evaluation of VaCARES is necessary to ensure that all birth defects, including NTDs, are being monitored efficiently and effectively. Using this evaluation, I plan to make recommendations for improving the quality, efficiency, and utility of the system.

I would like your input for this evaluation, and to facilitate responses, I have attached to this email a two-page feedback form. I hope you will feel free to let me know of any specific concerns or questions you want to have addressed by the evaluation. Upon completion I will communicate the findings of the evaluation to you so you can see how your contribution has helped VaCARES continue to improve its surveillance and education services. I look forward to working with you.

Thank you for your assistance.

Sincerely,

Caroline C. Stampfel
CSTE Fellow

VaCARES Surveillance Evaluation Stakeholder Survey – November 2005

1) How often do you have to deal with issues surrounding birth defects prevention and education? (Check only one)

__Every day

__1 to 4 times a week

__Less than once a week

__Less than once a month

__Less than once a year

__Other ________________

2) Place the following attributes of the VaCARES birth defects surveillance system in order from most important (1) to least important (4).

___ **Data quality** - validity of data recorded by the system.

___ **Representativeness** - the system gives an accurate picture of the occurrence of birth defects across all areas of Virginia.

___ **Sensitivity** – How many cases of birth defects has the system identified out of all who actually have birth defects.

___ **Stability** - reliability and availability of the public health surveillance system. Reliability refers to the ability to collect, manage, and provide data without failure. Availability refers to the ability to be operational when it is needed.

3) Do you get requests for birth defects information that cannot be addressed by the current VaCARES birth defects surveillance system?

___Yes

___No

___Not Sure

4) Which of the following do you think is most important to include in the reports from VaCARES? (Check only one)

__Birth defects by race and ethnicity

__Birth defects by health district

__Birth defects by county and city

__Birth defects by timing and quality of prenatal care

__Birth defects by other category________________ (please specify)

5) Which category of birth defect(s) is of greatest concern to health department staff and the public in your health district?

__Cardiovascular anomalies

__Clubfoot

__Down Syndrome

__Neural tube defects (anencephaly, spina bifida)

__Other defect _____________________(please specify)

6) What do you think VaCARES, as a statewide surveillance system, can do to assist your health district?

7) What can VaCARES do to improve education about birth defects in your district?

Virginia Congenital Anomalies Reporting and Education System
2005 Annual Report for Hospitals

Augusta Medical Center

Approximately 5% of children are born with birth defects, which are the leading cause of infant mortality. The Virginia Congenital Anomalies Reporting and Education System (VaCARES) was established and mandated by the Code of Virginia to collect information on infants and children diagnosed with birth defects. This information is used to address causes of birth defects, opportunities for prevention, and support for children and families. All hospitals in Virginia are required to report any child less than 24 months of age with a specified ICD-9 diagnosis code to the Virginia Department of Health. The expectation is that the report will be submitted through the Virginia Infant Screening and Infant Tracking System (VISITS) within 30 days of discharge.

Below you will find a summary of your hospital's reporting to VaCARES in 2004 and 2005. On the reverse is a brief comparison with other hospitals in Virginia.

2004 Reporting Summary

- Births at your hospital in 2004: **1097**
- Infants with birth defects discharged in 2004 and reported between 1/1/04 and 1/31/05: **0**
- Additional infants with birth defects discharged in 2004 reported between 2/1/05 and 1/31/06: **50**
- Total infants reported to VaCARES for 2004: **50**
- Percentage of births reported to VaCARES: **4.6%**

2005 Reporting Summary

- Births at your hospital in 2005 (preliminary): **1051**
- Infants with birth defects discharged in 2005 and reported between 1/1/05 and 1/31/06: **40**
- Percentage of births reported to VaCARES: **3.8%**
- Number of days between discharge and report to VaCARES: **126.5**

Feedback

Thank you for your commitment to VaCARES.
Please make sure to report all eligible infants. Please work on reporting in a timely manner.

Important Notes

1. Infants with addresses outside of Virginia are still eligible for VaCARES. Please ensure that all infants with birth defect ICD-9 codes are being reported.
2. VISITS is in the process of being redesigned. The new database should be available within the next two years. If you have ideas to make the system easier to use, please contact Allison Schreiber (804-864-7719 or Allison.Schreiber@vdh.virginia.gov).

Comments or Questions? Please contact:
Allison Schreiber, MS, CGC
Genetic Counselor, VaCARES Quality Control
Virginia Department of Health
(804) 864-7719 or Allison.Schreiber@vdh.virginia.gov

Hospital Name	2004 Births	2004 VaCARES Reporting	Percentage of Births	2005 Births	2005 VaCARES Reporting	Percentage of Births	2005 Reporting Time
llegheny Regional Hospital	50	3	6.0%	0	0	N/A	N/A
ugusta Medical Center	1097	50	4.6%	1051	40	3.8%	126.5
edford Memorial Hospital	193	8	4.1%	191	13	6.8%	26.7
on Secours DePaul Medical Center	1116	48	4.3%	1069	17	1.6%	29.1
on Secours Mary Immaculate Hospital	1017	56	5.5%	887	83	9.4%	18.6
on Secours Maryview Hospital	1078	39	3.6%	863	21	2.4%	53.7
on Secours Memorial Regional Medical Center	1438	18	1.3%	1634	0	0.0%	N/A
on Secours St. Francis Medical Center	0	0	N/A	140	0	0.0%	N/A
on Secours St. Mary's Hospital	2663	143	5.4%	2824	126	4.5%	42.4
uchanan General Hospital	51	0	0.0%	0	1	N/A	46.4
Carilion Franklin Memorial Hospital	243	10	4.1%	261	11	4.2%	35.7
Carilion New River Valley Medical Center	844	54	6.4%	884	52	5.9%	32.4
Carilion Roanoke Community Hospital	3039	422	13.9%	3145	468	14.9%	36.7
Chesapeake General Hospital	3324	199	6.0%	3103	149	4.8%	3.9
Children's Hospital of Richmond	0	3	N/A	0	3	N/A	81.6
Children's Hospital of The King's Daughters	0	577	N/A	0	601	N/A	12.8
CJW Medical Center – Chippenham Campus	1553	104	6.7%	1448	66	4.6%	61.4
CJW Medical Center – Johnston Willis Campus	2427	110	4.5%	2332	139	6.0%	7.7
Clinch Valley Medical Center	434	7	1.6%	433	10	2.3%	14.5
Community Memorial Health Center	278	7	2.5%	267	4	1.5%	4.2
Culpeper Regional Hospital	432	37	8.6%	487	40	8.2%	43.5
Danville Regional Medical Center	1048	19	1.8%	1126	28	2.5%	78.7
Dewitt Army Community Hospital	931	75	8.1%	995	48	4.8%	56.2
Fauquier Hospital	585	50	8.5%	650	55	8.5%	20.2
Halifax Regional Health System	455	26	5.7%	454	28	6.2%	6.6
Henrico Doctors' Hospital	3688	285	7.7%	3618	281	7.8%	121.2
Inova Alexandria Hospital	3543	281	7.9%	3619	383	10.6%	24.4
Inova Fair Oaks Hospital	3603	191	5.3%	3164	189	6.0%	19.8
Inova Fairfax Hospital	11329	925	8.2%	11463	648	5.7%	113.8
John Randolph Medical Center	516	11	2.1%	434	17	3.9%	9.3
Johnston Memorial Hospital	611	52	8.5%	600	53	8.8%	26.5
Lake Taylor Transitional Care	0	0	N/A	0	2	N/A	25.1
Langley Air Force Base	882	40	4.5%	951	22	2.3%	34.3
Lee County Regional Medical Center	3	0	0.0%	0	0	N/A	N/A
Lewis-Gale Medical Center	795	39	4.9%	611	17	2.8%	18.1
Loudoun Hospital Center	2121	223	10.5%	2210	221	10.0%	60.8
Martha Jefferson Hospital	1693	110	6.5%	1661	114	6.9%	16.8
Mary Washington Hospital	3524	240	6.8%	3507	398	11.3%	11.4
Memorial Hospital of Martinsville	540	0	0.0%	477	0	0.0%	N/A
Montgomery Regional Hospital	653	20	3.1%	583	23	3.9%	5.7
Naval Regional Medical Center, Portsmouth	4230	3	0.1%	4121	1	0.0%	125.6
Norton Community Hospital	467	40	8.6%	446	17	3.8%	8.7
Obici Memorial Hospital	1111	66	5.9%	1150	48	4.2%	62.3
Potomac Hospital	2454	132	5.4%	2531	127	5.0%	78.8
Prince William Hospital	2451	220	9.0%	2485	173	7.0%	87.9
Pulaski Community Hospital	133	21	15.8%	135	10	7.4%	19.7
Rappahannock General Hospital	43	0	0.0%	0	0	N/A	N/A
Reston Hospital Center	2841	290	10.2%	2953	243	8.2%	63.9
Riverside Regional Medical Center	2936	71	2.4%	2920	137	4.7%	95.8
Rockingham Memorial Hospital	1870	22	1.2%	1765	12	0.7%	20.6
Russell County Medical Center	0	0	N/A	0	0	N/A	N/A
Sentara Careplex Hospital	1014	0	0.0%	996	17	1.7%	106.1
Sentara Leigh Hospital	2308	0	0.0%	2622	2	0.1%	100.6
Sentara Norfolk General Hospital	2774	293	10.6%	2791	109	3.9%	119.9
Sentara Virginia Beach General Hospital	2020	185	9.2%	1808	112	6.2%	4.8
Sentara Williamsburg Community Hospital	898	67	7.5%	890	21	2.4%	251.0
Shenandoah Memorial Hospital	240	0	0.0%	237	2	0.8%	7.0
Shore Memorial Hospital	522	26	5.0%	527	48	9.1%	17.4
Smyth County Community Hospital	255	0	0.0%	273	0	0.0%	N/A
Southampton Memorial Hospital	275	10	3.6%	301	11	3.7%	45.2
Southern Virginia Regional Medical Center	149	1	0.7%	0	0	N/A	N/A
Southside Community Hospital	387	3	0.8%	380	1	0.3%	51.6
Southside Regional Medical Center	1263	17	1.3%	1253	12	1.0%	38.0
Stonewall Jackson Hospital	278	18	6.5%	288	21	7.3%	39.8
Twin County Regional Hospital	370	1	0.3%	371	7	1.9%	44.9
University of Virginia Health System	1527	702	46.0%	1682	791	47.0%	11.2
Virginia Baptist Hospital	2292	205	8.9%	2359	216	9.2%	24.7
Virginia Commonwealth University Medical Center	2202	661	30.0%	1985	787	39.6%	62.2
Virginia Hospital Center - Arlington	3246	201	6.2%	3225	305	9.5%	46.2
Warren Memorial Hospital	277	3	1.1%	212	8	3.8%	4.2
Wellmont Lonesome Pine Hospital	245	2	0.8%	222	0	0.0%	N/A
Winchester Medical Center	2172	93	4.3%	2306	121	5.2%	35.0
Wythe County Community Hospital	229	5	2.2%	252	0	0.0%	N/A
Total	101276	7840	7.7%	100628	7730	7.7%	46.8

date

Re: [child's name] DOB:

Dear [name],

This letter is to provide you with information that may be helpful to you and your child. The enclosed guide tells about services that can help families who have a child with special health care needs.

You were sent this letter because the Virginia Department of Health was informed that you might have a child with a special heath care need. It is Virginia law that hospitals report any child under the age of 2 who has a birth defect to the Virginia Congenital Anomaly Reporting and Education System (VaCARES). No information that will identify you or your child will be used in any publication or report. The reports are used for the following reasons:

1. To study possible causes of birth defects.
2. To improve the treatment of birth defects.
3. To tell parents about helpful resources.

We make every effort to not contact parents who have experienced the death of a child. If this has happened, we apologize for sending this letter. Please accept our sympathy and let us know if this has happened.

We hope that this letter and the enclosed brochure are helpful. If you have questions, please call our toll free number, 1-800-523-4019. Someone is available to talk with you Monday thru Friday, from 8 a.m. to 4:30 p.m.

Sincerely,

Sharon K. Williams, MS, RN
Virginia Genetics Program Manager
Pediatric Screening and Genetic Services
Division of Child and Adolescent Health
Virginia Department of Health

Cc: [Primary Care Physician]

VaCARES: Virginia Congenital Anomalies Reporting and Education System Surveillance Methodology Evaluation Plan

Use the *Updated Guidelines for Evaluating Public Health Surveillance Systems: Recommendations From the Guidelines Working Group* published by the Centers for Disease Control and Prevention[1]. The steps for evaluating a surveillance system are outlined here with specific attention to how these steps relate to VaCARES.

1. Engage the Stakeholders in the Evaluation

1.1 Contacts may include:

- Virginia Department of Health staff who oversee the surveillance system in DCAH Pediatric Screening and Genetic Services, DWIH Folic Acid Program
- Representatives of the 35 Health Districts – Nurse Managers or Health Directors
- Virginia Genetics Advisory Committee – includes parents, physicians, and representatives from academia and the health department
- Virginia Council on Folic Acid
- March of Dimes
- National Birth Defects Prevention Network (for materials and guidelines for BD surveillance)
- Hospital data entry users

1.2 Contact may be in the form of:

- Letters or emails notifying the stakeholder of the evaluation
- Surveys
- Structured feedback forms
- Phone calls
- In-person interviews

2. Describe VaCARES

This step provides necessary background to understand why Virginia has a surveillance system for birth defects and how this system currently functions. The previous evaluation can be a starting point for future evaluations, and every effort should be made to use the most current statistics, cost estimates, and surveillance system information. The disease category "birth defects" encompasses a variety of conditions that vary widely in severity, cost, and preventability. When possible, statistics available for 1) neural tube defects (NTDs), 2) Down Syndrome, and 3) congenital heart defects should be reported because the data tend to be recent and plentiful.

2.1 Describe the public health importance of birth defects

Compare Virginia with published national rates or estimates, and use VaCARES published statistics.

a) Indices of frequency – prevalence, total number of birth defects, birth defect attributed mortality, and summary measures of population health status (QALYS)

b) Indices of severity of birth defects – bed-disability days, case-fatality ratio, hospitalization / disability rates.

c) Disparities associated with birth defects – racial/ethnic, disabilities, social

d) Costs associated with birth defects
e) Preventability (primary, secondary, and tertiary prevention)
f) Public Interest

2.2 Describe the purpose and operation of VaCARES
a) Planned uses of the data from VaCARES
b) Case definition for birth defects: ICD9-CM codes that qualify for VaCARES
c) Legal authority for data collection:
- The legal authority for data collection and the development of VaCARES was mandated in 1985 and amended in 1986 in the *Code of Virginia*, §32.1-69.1.

d) Describe the organizational location of VaCARES
e) Describe the relevant political, administrative, geographic, or social context in which the evaluation of VaCARES will be done.
f) Describe the level of integration with other systems
- VISITS, integration with Newborn Screening, Early Hearing Screening, Lead Screening
- Integration with programs such as Folic Acid Campaign

g) Draw a flow chart of the system
h) Describe the components of VaCARES, including the following:
- Population under surveillance (Virginia Birth Population)
- Period of time of the data collection
- What data are collected and how
- Reporting sources – hospitals and regional genetics centers
- Data management policies, compliance with standards fir formats and coding
- Data analysis plans and dissemination plans
- Privacy policies, confidentiality policies, and system security
- Records management program

2.3 Describe the resources used to operate VaCARES
a) Funding sources
b) Personnel requirements
c) Other resources

3. Focus the Evaluation Design

3.1 Determine the specific purpose of the evaluation.
3.2 Determine which stakeholders will receive the findings and recommendations of the evaluation.
3.3 Specify the questions that will be answered by the evaluation
- Use stakeholder contacts to narrow down the focus of the evaluation. Not all of the system attributes need to be explored in detail. Find out if any of the components have been addressed recently and can be incorporated into the evaluation.

3.4 Determine the standards for assessing the performance of the system:

- National Birth Defects Prevention Network Guidelines for Conducting Birth Defects Surveillance
- CDC's Updated Guidelines for Evaluating Public Health Surveillance Systems
- Trust for America's Health: Birth Defects Tracking and Prevention: Too Many States Are Not Making the Grade
- Compare VaCARES to other birth defects surveillance systems, such as Illinois.

4. Gather Credible Evidence Regarding the Performance of the Surveillance System

4.1 Indicate the level of usefulness of VaCARES.
  a) Does VaCARES address its stated objectives?
  b) What is VaCARES effect on policy decisions and disease-control programs?

4.2 Describe each surveillance system attribute as it applies to VaCARES:

- a) Simplicity – Structure and ease of operation should be as simple as possible while still meeting objectives.
  - a. Evaluate system for linking records to birth certificates and death certificates.
  - b. Evaluate procedures for amending records and de-duplicating the database.
  - c. Look at confirmed vs. provisional defects in system.
  - d. Evaluate infrastructure and integration of VaCARES with other health department systems.

- b) Flexibility – Able to adapt to changing information needs or operating concerns with little additional time, personnel, or allocated funds.
  - a. Examine some scenarios in which information needs change and discuss with staff impact of these changes.
  - b. Examine the past adaptations of the system, evaluate the consequences, and predict how the current system would react.

- c) Data quality – Completeness and validity of data recorded by the system.
  - a. Examine the number of responses that are 'unknown' and blank.
  - b. Evaluate the utility of collecting birth defects beyond those recommended by NBDPN.
  - c. Evaluate mandatory and optional designations for data entry items.

- d) Acceptability – Willingness of persons and organizations to participate in the surveillance system.
  - a. Conduct user satisfaction survey.
  - b. Calculate hospital reporting rate.
  - c. Evaluate data entry time.

- e) Sensitivity – Proportion of cases of a disease detected by the surveillance system. This also includes the ability to detect "outbreaks" and monitor changes in disease patterns over time.

   a. Calculate expected rate for Virginia and compare to what is detected by the surveillance system by health district.
   b. Evaluate how well VaCARES identifies deceased children.

f) Predictive Value Positive – The proportion of reported cases that actually have the health–related event under surveillance.
   a. Make comparison to birth records. We expect birth defects to be underreported on birth certificates.

g) Representativeness – The system accurately describes the occurrence of a health-related event over time and its distribution in the population by place and person.
   a. Calculate expected rate for Virginia and compare to what is detected by the surveillance system by health district.
   b. Explore using GIS to evaluate representativeness of VaCARES.

h) Timeliness – The speed between steps in the surveillance. Includes the amount of time between event and event recognition, between event recognition and event reporting, between reporting and action, between reporting to public health agency and feedback to stakeholders.
   a. Look at time between diagnosis and reporting to VaCARES.
   b. Look at time between completion of birth cohort and reporting time.

i) Stability – Reliability and availability of the public health surveillance system. Reliability refers to the ability to collect, manage, and provide data without failure. Availability refers to the ability to be operational when it is needed.
   a. Evaluate financial support for VaCARES and loss of funding scenarios.
   b. Evaluate staffing support for VaCARES.

5. Justify and State Conclusions, and Make Recommendations

5.1 State conclusions with justification from gathered evidence and analyses.
   a) Does VaCARES address a relevant public health problem?
   b) Is VaCARES meeting its objectives?
   c) Has VaCARES been updated to meet any previous evaluation recommendations? Why or why not?
5.2 Make recommendations based on conclusions, taking into account the impact of recommendations.

6. Ensure Use of Evaluation Findings and Share Lessons Learned

Findings should be presented to stakeholders in the most appropriate format. For example, VaCARES staff should receive the full text of the evaluation, but stakeholders that have less technical knowledge, such as parents, might require an executive summary or brief.

Dissemination activities may include:

- Submitting evaluation as an abstract to NBDPN's meeting or another national meeting or conference (MCH Epidemiology, APHA, etc)
- Submitting evaluation to a journal for publication (Public Health Reports published a birth defects surveillance evaluation in New York State 2001).
- A Polycom video conference with district health directors and/or nurse managers
- Issue brief / executive summary to all stakeholders contacted
- Posting on Virginia Department of Health website

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[1] Centers for Disease Control and Prevention. Updated Guidelines for Evaluating Public Health Surveillance Systems: Recommendations From the Guidelines Working Group. MMWR 2001;50(No. RR-13): [4-25].

PEDIATRIC SCREENING AND GENETIC SERVICES
Testing, Tracking, Treating

Facts about
VaCARES: Virginia Congenital Anomalies Reporting and Education System

Birth Defects in Virginia

- Each year in Virginia, about 4600 infants are born with birth defects, of which about 270 will die before the age of 2.
- In Virginia, as elsewhere in the United States, birth defects are the leading cause of death in the first year of life.
- Birth defects may be due to genetic or environmental factors, but the cause of most birth defects is unknown.

Mission of VaCARES

- Collect data to evaluate possible causes of birth defects.
- Improve the diagnosis and treatment of birth defects.
- Establish a mechanism for informing parents of children identified as having birth defects and their physicians about available health resources.

Legislation and Administration

- The development of the statewide VaCARES was mandated in 1985 and amended in 1986 in the *Code of Virginia,* § 32.1-69.1.
- VaCARES is managed by the Virginia Department of Health, Division of Child and Adolescent Health, Pediatric Screening and Genetic Services.
- Expert consultation is provided by the Virginia Commonwealth University, Department of Human Genetics.
- The Virginia Genetic Advisory Committee, established under the *Code of Virginia,* § 32.1-69.1, assists in the ongoing improvement of VaCARES.

Virginia Department of Health
Division of Child and Adolescent Health
Pediatric Screening and Genetic Services
Virginia Genetics Program
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www.vdh.state.va.us

Data Collection

- Every hospital, as mandated by *Code of Virginia,* § 32.1-123, reports to the Virginia Department of Health any child under the age of 2 years diagnosed as having a birth defect.
- Eighty-six different categories of structural, functional, or biochemical abnormalities are required to be reported.
- Data obtained from Virginia Newborn Screening Services are also entered into the VaCARES database.
- Additional data from birth and death certificates filed with the State Registrar of Vital Records are added to VaCARES.

Potential Uses of VaCARES Data

- Identify birth defect trends and state locations with high rates of birth defects.
- Evaluate the success of prevention programs aimed at reducing the incidence of birth defects in Virginia.
- Assess the impact of birth defects on families and communities.
- Connect children with birth defects to resources in their communities.
- Investigate community concerns about birth defects.
- Enhance public awareness and knowledge about birth defects.
- Provide information to scientists that may lead to a better understanding of birth defects.

Data Limitations

- Data are based on passive reporting, which means that facilities (i.e., hospitals) are responsible for identifying and reporting cases of birth defects. Facilities may vary in their completeness and timeliness of reporting.
- Some facilities may report children with a birth defect that is later ruled out, resulting in an overestimate of birth defects.
- The VaCARES program monitors birth defects reported in Virginia in children under the age of 2. Children diagnosed with birth defects 2 or more years after birth would not be included in the report, resulting in an underestimate of birth defects.
- Children diagnosed and treated in other states may be missed.
- In some cases, hospitals may misdiagnose or miscode birth defects.
- Data collected does not include miscarriages, pregnancy losses, and stillbirths associated with birth defects.

Confidentiality

- The confidentiality of the information collected for VaCARES is maintained by the Virginia Department of Health and protected by the *Code of Virginia,* § 32.1-69.2.

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PEDIATRIC SCREENING AND GENETIC SERVICES
Testing, Tracking, Treating

Facts about

Cleft Lip and Cleft Palate

What are cleft lip and cleft palate?

A cleft is a "split." Cleft lip and cleft palate are types of oral-facial clefts.

The lip and palate (hard and soft tissue forming the roof of the mouth) begin developing in separate parts in a growing baby. These parts fuse together to form the complete lip and palate between weeks 5 and 10 of the baby's growth. A cleft forms when the sides of the lip or palate do not join correctly as the baby develops.

A cleft lip is, therefore, a split of the two sides of the lip that can include the gums and bones of the jaw. A cleft palate is an opening or split in the roof of the mouth. Cleft lip and cleft palate can occur on one side of the lip or palate (unilateral) or on both sides (bilateral). A cleft may occur only in the lip or only in the palate, or it may occur in both the lip and palate simultaneously.

What types of problems occur with cleft lip and/or cleft palate?

Feeding problems (e.g., taking in too much air while eating, feeding slowly, milk passing through the nose) do not usually occur in children with a cleft lip alone but are common in children with cleft palate. Ear infections due to a build up of fluid in the ear and speech problems can also occur. Depending on the extent of the cleft, problems may also occur with a child's teeth such as missing, extra, or crooked teeth. Between 15 to 50 percent of children with cleft lip and/or cleft palate have other major birth defects.

How common are cleft lip and/or cleft palate?

Cleft lip and cleft palate are the fourth most common birth defect in the United States, occurring in about 5,000 births yearly. In Virginia about 120 children are born yearly with cleft lip and/or cleft palate. Greater than 70% of babies with cleft lip will also have cleft palate.

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What causes cleft lip and/or cleft palate?

It is believed that most cleft lip and/or cleft palate occur due to a combination of environmental and genetic factors. Environmental factors may include maternal illness, medications, maternal smoking, and infections. Most children with cleft lip and/or cleft palate are born into families with no family history of cleft lip and/or cleft palate. Although some families have more than one person with cleft lip and/or palate, clefting does not occur in these families in a set pattern. Parents of a child with cleft lip and/or palate have a higher chance of having another child with cleft lip and/or palate. This chance is about 2 to 8 in 100. For parents who themselves had a cleft lip and/or cleft palate, the chance of having a child with a cleft lip and/or cleft palate is about 4 to 6 in 100.

Cleft lip and/or cleft palate can be the only birth defects a child has (an isolated finding), but they can also occur with other birth defects as part of a syndrome (a collection of findings). The way in which a syndrome is passed through the family is specific to the given syndrome. A genetic counselor or geneticist can help you to determine the risks for your family and situation.

How are cleft lip and/or cleft palate treated?

Surgery is typically done to close the cleft during infancy. Additional surgeries may be needed during childhood and adolescence. Special positioning of the child during feeding or special bottles may help with feeding problems. Children with clefting involving only the lip are likely to have fewer problems. If the clefting involves the gums and/or the palate, the child may need dental care to address the growth of the teeth and may need speech therapy. To prevent hearing loss and ear infection, medications may be given or surgery may be done to drain any fluid in the ear. Often a team of specialists manages the care of a child with cleft lip and/or cleft palate. Treatment may extend over a period of several years.

Where can I go for more information about cleft lip and/or palate?

Cleft Palate Foundation
www.cleftline.org
1-800-24-CLEFT (1-800-242-5338)

March of Dimes Birth Defects Foundation
www.modimes.org
1-888-MODIMES (1-888-663-4637)

Infant and Toddler Connection of Virginia (Early Intervention Services)
www.infantva.org
1-800-234-1448

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Datos sobre
la hendidura del labio y del paladar

¿Qué es la hendidura del labio y del paladar?

Una hendidura es una "abertura". La hendidura del labio y del paladar es un tipo de abertura orofacial (en la boca y la cara).

El labio y el paladar (el tejido duro y blando que forma la parte superior de la boca) comienzan a desarrollarse como partes separadas en un bebé en crecimiento. Estas partes se unen para formar la estructura completa de los labios y del paladar de 5 a 10 semanas después de iniciarse el período de crecimiento del bebé. Se forma una hendidura cuando los lados del labio o del paladar no se unen correctamente a medida que se desarrolla el bebé.

Por tanto, una hendidura del labio es una abertura de ambos lados del labio que puede extenderse a las encías y a los huesos de la mandíbula. Una hendidura del paladar es una grieta o abertura en la parte superior de la boca. La hendidura del labio y del paladar puede ocurrir a un lado del labio o del paladar (hendidura unilateral) o a ambos lados (hendidura bilateral). Una hendidura puede ocurrir solo en el labio o solo en el paladar o puede presentarse simultáneamente en el labio y el paladar.

¿Qué tipos de problemas causa la hendidura del labio y/o del paladar?

Los problemas de alimentación (por ejemplo, aspiración de demasiado aire al comer, alimentación lenta, paso de leche a través de la nariz) no suelen ocurrir únicamente en los niños con hendidura del labio, sino que son comunes también en los niños con hendidura del paladar. Además pueden ocurrir infecciones del oído por acumulación de líquido y problemas del habla. Según el tamaño de la hendidura, también pueden presentarse problemas de dentición (crecimiento de los dientes) del niño, por ejemplo, algunos dientes pueden faltar o crecer torcidos o el número de dientes puede ser mayor número del normal. Entre 15 y 50% de los niños con hendidura del labio y/o del paladar tienen otros defectos congénitos (que se presentan al nacer) graves.

¿Qué tan común es la hendidura del labio y/o del paladar?

La hendidura del labio y del paladar ocupa el cuarto lugar entre los defectos congénitos más comunes en los

Estados Unidos y ocurre en unos 5.000 nacimientos al año. En Virginia nacen anualmente unos 120 niños con hendidura del labio y/o del paladar. Más de 70% de los niños con hendidura del labio también tendrán hendidura del paladar.

¿Qué causa la hendidura del labio y/o del paladar?

Se cree que la mayoría de los casos de hendidura del labio y/o del paladar ocurren por un conjunto de factores ambientales y genéticos. Los factores ambientales pueden incluir enfermedad materna, uso de medicamentos, hábito de fumar de la madre e infecciones. Casi todos los niños con hendidura del labio y/o del paladar pertenecen a familias sin antecedentes de ese defecto. Aunque algunas familias tienen más de una persona con hendidura del labio y/o del paladar, ese defecto no ocurre en esas familias según un patrón establecido. Los padres de un niño con hendidura del labio y/o del paladar tienen mayor posibilidad de tener otro con el mismo defecto. Esa posibilidad es de alrededor de 2 a 8 en 100. Cuando los padres tienen hendidura del labio y/o del paladar, su posibilidad de tener un hijo con el mismo defecto es aproximadamente de 4 a 5 en 100.

La hendidura del labio y/o del paladar puede ser el único defecto congénito que tiene un niño (un hallazgo aislado), pero también puede ocurrir junto con otros defectos congénitos como parte de un síndrome (un conjunto de hallazgos). La forma en que se hereda un síndrome en la familia es específica de ese síndrome Un asesor en genética o un genetista puede ayudarle a determinar los posibles riesgos para su familia y su situación.

¿Cómo se trata la hendidura del labio y/o del paladar?

Típicamente, se realiza una cirugía para cerrar la hendidura en la infancia. Es posible que se necesiten otras operaciones durante la niñez y la adolescencia. La colocación del niño en una posición particular durante la alimentación o el uso de biberones especiales pueden ayudar a resolver los problemas de la alimentación. Los niños con hendidura solamente del labio pueden tener menos problemas. Si la hendidura abarca las encías y/o el paladar, el niño puede necesitar atención dental para resolver los problemas de dentición y terapia del lenguaje. Para evitar la pérdida de la audición y la infección de los oídos, se pueden dar medicamentos o realizar una operación para drenar cualquier líquido del oído. A menudo se necesita un equipo de especialistas para atender al niño con hendidura del labio y/o del paladar. El tratamiento puede durar varios años.

¿Dónde puedo obtener más información sobre la hendidura del labio y/o del paladar?

Cleft Palate Foundation
[Fundación para la Hendidura del Paladar]
www.cleftline.org
1-800-24-CLEFT (1-800-242-5338)

March of Dimes Birth Defects Foundation
[Fundación de Niños con Defectos Congénitos March of Dimes]
www.modimes.org
1-888-MODIMES (1-888-663-4637)

Infant and Toddler Connection of Virginia (Early Intervention Services)
[Conexión de Virginia para Lactantes y Niños Pequeños (Servicios de Intervención Temprana)]
www.infantva.org
1-800-234-1448

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Appendix B – Tables

Table 1 - Code of Virginia §32.1-69.1

Virginia Congenital Anomalies Reporting and Education System
A. In order to collect data to evaluate the possible causes of birth defects, improve the diagnosis and treatment of birth defects and establish a mechanism for informing the parents of children identified as having birth defects and their physicians about the health resources available to aid such children, the Commissioner shall establish and maintain a Virginia Congenital Anomalies Reporting and Education System using data from birth certificates filed with the State Registrar of Vital Records and data obtained from hospital medical records. The chief administrative officer of every hospital, as defined in § 32.1-123, shall make or cause to be made a report to the Commissioner of any person under two years of age diagnosed as having a congenital anomaly. This report shall include information on the parent's service in Vietnam, the duration of the service and possible exposure to Agent Orange through its development, testing or use. The Commissioner may appoint an advisory committee to assist in the design and implementation of this reporting and education system with representation from relevant groups including, but not limited to, physicians, geneticists, personnel of appropriate state agencies, persons with disabilities and the parents of children with disabilities.
B. With the assistance of the advisory committee, the Board shall promulgate such regulations as may be necessary to implement this reporting and education system. These regulations may include determinations of specific genetic disorders to be monitored, the scope of the information to be collected, appropriate mechanisms for follow-up, relationships between the reporting and education system and other agencies and mechanisms for review and evaluation of the activities of the system. The reporting and education system may collect the name, address, sex, race, and any other information, determined to be pertinent by the Board, regarding persons reported to have birth defects. (1985, c. 273; 1986, c. 136; 1988, cc. 459, 843; 1994, c. 854.)

Table 2: ICD-9-CM Diagnosis Code Groups Reportable to VaCARES

ICD-9-CM4 Code	Anomaly
042	Human immunodeficiency virus disease
090	Congenital syphilis
189-191	Wilms tumor, retinoblastoma, other congenital neoplasms
237-259	Endocrine disorders
270-275, 277-279	Metabolic and immune disorders
282-284, 286	Specified anemias and coagulation defects
330-343, 352-359	Nervous system disorders
362-363	Retinal Disorders
369	Blindness
389	Hearing loss
425-427	Cardiomyopathy and conduction disorders
524	Dentofacial anomalies
740-759	Congenital anomalies
760.7	Noxious influences affecting fetus via placenta or breast milk
771.0-771.2	Selected congenital infections

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Table 3: Selected Birth Outcomes by Race/Ethnicity, Virginia, 2004[a].

	All Races/ Ethnicities		White Non-Hispanic		Black Non-Hispanic		Hispanic Any Race	
Birth Outcome/ Risk Factor	N	Rate[b] or %	N	Rate[b] or %	N	Rate[b] or %	N	Rate[b] or %
Live Births	103,830	--	62,662	--	22,111	--	11,658	--
Infant Deaths (< 1 year)	768	7.4	358	5.7	312	14.1	66	5.7
Early Infant Deaths (<1 day)	335	3.2	136	2.2	142	6.4	39	3.3
Neonatal Deaths (<28 days)	518	5.0	232	3.7	207	9.4	52	4.5
Postneonatal Deaths (28-364 days)	250	2.4	126	2.0	105	4.7	14	1.2
Low Birthweight (<2500g)	8,674	8.4%	4,451	7.1%	2,853	12.9%	755	6.5%
Very Low Birthweight (<1500g)	1,699	1.6%	843	1.3%	653	3.0%	142	1.2%
1st Trimester Entry Into Prenatal Care	87,932	85.9%	56,002	89.4%	17,431	78.8%	8,344	71.6%

[a] Source: Virginia Center for Health Statistics compiled by the Office of Family Health Services

[b] Rate per 1,000 live births

Table 4: Variables Reported by Hospitals and Vital Records

	Required from Hospital Users	Not Required but Collected	Collected from Vital Records
Unique ID	+		+
Sex	+		+
Infant's Name: First	+		+
Middle		+	+
Last	+		+
Suffix		+	+
Source of Report	+		+
Medical Record Number(s)	+		+
Vital Record Certificate Number		+	+
Place of Pregnancy Outcome	+		+
Pregnancy Outcome			+
Birth Weight			+
Plurality		+	+
Gestational Age			+
Diagnosis code	+		+
Name of Responsible Party	+		+
Address of Responsible Party	+		+
Telephone of Responsible Party		+	
Mother's Date of Birth			+
Mother's Race			+
Mother's Ethnicity			+
Mother's Name: First	+		+
Middle		+	+
Last	+		+
Mother's Residence at Time of Pregnancy Outcome			
Street Address		+	+
City		+	+
County		+	+
State		+	+
Zip Code		+	+

Table 5: State and Federal Laws and Regulations

Reporting and Education Requirements	
Code of Virginia § 32.1-69.1 Virginia Congenital Anomalies Reporting and Education System.	• Authorizes the Health Commissioner to establish VaCARES • Requires hospitals to report birth defects to the health department
Code of Virginia § 32.1-69.1:1 Dissemination of information regarding birth defects.	• Health Commissioner will develop folic acid publications for prevention of birth defects and distribute to health care providers, medical facilities, and health departments.
Social Security Act, Title V Sections 505 and 506	• Outline the HRSA MCHB Block Grant state allocations • Reporting requirements to ensure proper use of the grant.
Confidentiality	
Code of Virginia § 32.1-69.2 Confidentiality of records; publication; authority of Commissioner to contact parents and physicians.	• Health Commissioner will keep confidential all health records and will only make public de-identified aggregate statistics. • Health Commissioner may contact the parents of children with birth defects and their physicians in order to collect relevant data and provide information about resources.
Code of Virginia § 32.1-127.1:03 Health Records Privacy	• Recognizes an individual's right of privacy in the content of his or her health records • Defines instances when they records be released to the individual or other entity.
Code of Virginia § 32.1-127.1:04 Use or disclosure of certain protected health information required	• Department of Health and other health agencies shall establish a secure system for sharing protected health information necessary for prevention and control of disease, injury or disability.
Security Standards	
VDH ITRM Policies and Procedures	• Establishes a framework for the development and governance of Commonwealth of Virginia Information Technology Resource Management (ITRM) Policies,

	Standards, and Guidelines as well as other Policies, Standards, and Guidelines involved with information technology related issues. • The management of information technology (IT) resources requires the establishment and control of a set of documents that convey purpose, direction, and required activities.
COV ITRM Standard SEC2001-01.1	• Information technology security standard for Virginia state agencies, carried out by VITA, the Virginia Information Technologies Agency. • Includes 13 components that provide a framework to enable secure communications and the appropriate protection of information resources within the Commonwealth and provide the basis for designing the Agency's security program and safeguards.

Table 6: Frequency of Defect Status 2003-2005

Defect Status	Frequency	Percent
Confirmed	37802	95.45
Provisional	994	2.51
Missing	491	1.24
Unknown	315	0.8

Table 7: Fields Where Percent Completed is Less than 95%, 2003-2005

Field Name	% Completed
Medical Record Number	24.40%
Patient Control Number	0.03%
Birth Certificate Number	0.04%
Vitals Birth Certificate Number	0.01%
Death Certificate Number*	0.90%
Screening Date	0.20%
First Name (Child)	86.30%
Birth Hospital	85.40%
Plurality	4.10%
Mother's Maiden Name	5.90%
Ethnicity2 (Hispanic/non-Hispanic)	9.00%
Hispanic	0.01%
Race	0.01%
Mother's First Name**	94.20%
Mother's Last Name**	94.20%

*Of those listed as deceased
**Borderline

Table 8: Values for Race Variable From VISITS-Provided Reference Table

Value	Definition
1	White/Caucasian
2	Black or African American
3	Amer.Ind./Alaskan Native
8	Native Hawaiian or Other Pacific Islander
9	Unknown
10	Other
12	More than One Race Reported
13	Hispanic
406	Other Asian or Pacific Islander
407	Others
409	Unknown, not stated, unclassified
410	Filipino
412	Chinese
417	Japanese
11	Asian

Table 9: True Definitions for Variable Ethnicity (Race)

Value	Definition
1	White / Caucasian
2	Black / African American
3	American Indian / Alaskan Native
4	Chinese
5	Japanese
6	Hawaiian
7	Filipino
8	Other Asian Pacific Islander (?)
9	Unknown
10	Other
11	Asian
12	Asian Indian

Table 10: Values for Ethnicity From VISITS-Provided Reference Table

1	Afghan	22	Finnish	42	Latvian	61	Syrian
2	African	23	French	43	Lebanese	62	Taiwanese
3	Albanian	24	German	44	Lithuanian	63	Thai
4	American	25	Greek	45	Mexican	64	Ukrainian
5	American Indian	26	Guamanian	46	Mongolian	65	Vietnamese
6	Arab	27	Guyanese	47	Nicaraguan	66	Welsh
7	Armenian	28	Haitian	48	Norwegian	67	Yugoslavian
8	Austrian	29	Hawaiian	68	Others	69	UNKNOWN
9	Belgian	30	Hispanic	49	Pakistani	99	Cambodian
11	Canadian	31	Hmong	50	Persian	411	Latin American
12	Chilean	32	Hungarian	51	Polish	405	Unknown Not stated Unclassified
13	Chinese	33	Indian	52	Portuguese		
14	Colombian	34	Iranian	53	Puerto Rican		
15	Croatian	35	Irish	54	Romanian		
16	Cuban	36	Israeli	55	Russian		
17	Czech	37	Italian	56	Samoan		
18	Danish	38	Jamaican	57	Scottish		
19	Dutch	39	Japanese	58	Spanish		
20	English	40	Korean	59	Swedish		
21	Filipino	41	Latin Amer.	60	Swiss		

Table 11: True Definitions for Variable Ethnicity2 (Ethnicity)

Value	Definition
1	Hispanic
2	Non-Hispanic

Table 12: Sensitivity and Positive Predictive Value 2X2 Table

		Birth Defect Present in Virginia Child age 0-2		
		YES	**NO**	
Birth Defect Detected by VaCARES	**YES**	True positive **A**	False positive **B**	**A+B**
	NO	False positive **C**	True Negative **D**	**C+D**
		A+C	**B+D**	**Total**

Sensitivity = A / (A+C)

Positive Predictive Value = A / (A+B)

Table 13: VaCARES to Birth Certificate Birth Defects Crosswalk Table

Birth Certificate Birth Defects Categories	Corresponding ICD-9-CM codes from VaCARES
01-Anencephalus	740-740.2
02-Spina bifida / Meningocele	741-741.93
03-Hydrocephalus	742.3
04-Microcephalus	742.1
05-Other central nervous system anomalies	742 or 742.0 or 742.2 or 742.4 or 742.5-742.9
06-Heart malformations	745.0-746.9
07-Other circulatory/ respiratory anomalies	747.0-748.9
08-Rectal atresia/ stenosis	751.2
09-Tracheo-esophageal fistula/ esophageal atresia	750.3
10-Omphalocele/ Gastroschisis	756.7 or 756.79
11-Other gastrointestinal anomalies	750 - 751.9
12-Malformed genitalia	752.0-752.9
13-Renal Agenesis	753, 753.0
14-Other urogenital anomalies	753.1-753.9
15-Cleft lip / palate	749-749.25
16-Polydactyly / syndactyly / adactyly	755.0-755.14
17-Club foot	754.5-754.79
18-Diaphragmatic hernia	756.6
19-Other musculoskeletal/ integumental anomalies	754 -754.44 or 754.8-754.89 or 755.2-756.59 or 756.8-757.9
20-Down’s syndrome	758.0
21-Other chromosomal anomalies	758.1-758.9
22-other	759, 759.8, 759.9

Table 14: NBDPN Birth Defects Counts and Rates 1998-2002

(Rates per 10,000 live births)

VaCARES Reportable Defect	Race / Ethnicity						
	Non-Hispanic White	Non-Hispanic Black or African	Hispanic	Asian or Pacific Islander	American Indian or Alaskan Native	Other / Unknown	**Total**
Amniotic Bands	1	0	0	0	0	0	**1**
	0.07	0.00	0.00	0.00	0.00		**0.05**
Anencephalus	15	2	2	0	0	1	**20**
	0.47	0.18	0.52	0.00	0.00		**0.40**
Aniridia	2	0	0	0	0	1	**3**
	0.06	0.00	0.00	0.00	0.00		**0.06**
Anophthalmia / microphthalmia	7	4	1	0	0	0	**12**
	0.22	0.37	0.26	0.00	0.00		**0.24**
Anotia / microtia	6	2	1	1	0	2	**12**
	0.19	0.18	0.26	0.44	0.00		**0.24**
Aortic Valve Stenosis	16	5	1	3	0	15	**40**
	0.50	0.46	0.26	1.32	0.00		**0.80**
Atrial septal defect	635	175	83	47	0	858	**1,798**
	19.81	16.02	21.53	20.70	0.00		**35.94**
Biliary atresia	11	6	1	2	0	11	**31**
	0.34	0.55	0.26	0.88	0.00		**0.62**
Bladder exstrophy	3	0	0	0	0	1	**4**
	0.09	0.00	0.00	0.00	0.00		**0.08**
Choanal atresia	25	7	1	1	0	18	**52**
	0.78	0.64	0.26	0.44	0.00		**1.04**
Cleft lip with and without cleft palate	180	26	20	11	0	3	**240**
	5.62	2.38	5.19	4.84	0.00		**4.80**
Cleft palate without cleft lip	97	24	4	6	0	35	**166**
	3.03	2.20	1.04	2.64	0.00		**3.32**
Coarction of aorta	104	16	4	6	0	66	**196**
	3.24	1.46	1.04	2.64	0.00		**3.92**
Common truncus	10	7	1	2	0	15	**35**
	0.31	0.64	0.26	0.88	0.00		**0.70**
Congenital cataract	14	5	1	0	0	12	**32**
	0.44	0.46	0.26	0.00	0.00		**0.64**
Congenital hip dislocation	236	17	12	6	0	91	**362**
	7.36	1.56	3.11	2.64	0.00		**7.24**
Diaphragmatic hernia	46	14	4	4	0	23	**91**
	1.44	1.28	1.04	1.76	0.00		**1.82**
Down syndrome (trisomy 21)	228	64	24	14	1	137	**468**
	7.11	5.86	6.23	6.16	15.11		**9.36**
Ebstein's anomaly	9	0	0	0	0	9	**18**
	0.28	0.00	0.00	0.00	0.00		**0.36**

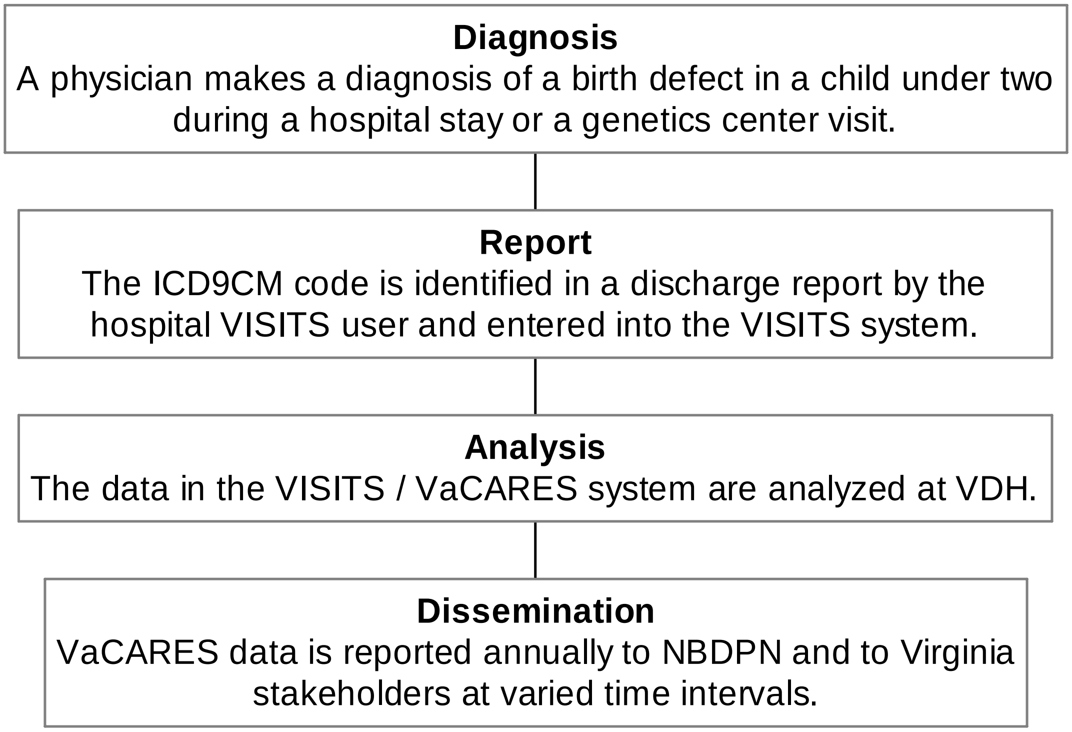
Encephalocele	6	5	0	0	0	6	**17**
	0.19	0.46	0.00	0.00	0.00		**0.34**
Endocardial cushion defect	56	18	1	0	0	44	**119**
	1.75	1.65	0.26	0.00	0.00		**2.38**
Esophageal atresia / tracheoesophageal fistula	55	9	4	1	0	25	**94**
	1.72	0.82	1.04	0.44	0.00		**1.88**
Fetus or newborn affected by maternal alcohol use	16	23	1	0	1	10	**51**
	0.50	2.11	0.26	0.00	15.11		**1.02**
Hirschprung's disease (congenital megacolon)	34	22	3	0	0	35	**94**
	1.06	2.01	0.78	0.00	0.00		**1.88**
Hydrocephalus without Spina Bifida	75	51	11	3	0	30	**170**
	2.34	1.67	2.85	1.32	0.00		**3.40**
Hypoplastic left heart syndrome	40	10	0	1	0	26	**77**
	1.25	0.92	0.00	0.44	0.00		**1.54**
Hypospadias and Epispadias	244	78	6	6	1	170	**630**
	7.61	7.14	1.56	2.64	15.11		**12.59**
Microcephalus	52	35	6	7	0	45	**145**
	1.62	3.20	1.56	3.08	0.00		**2.90**
Obstructive genitourinary defect	97	41	7	6	0	9	**160**
	3.03	3.75	1.82	2.64	0.00		**3.20**
Patent ductus arteriosus	916	462	102	70	4	1,036	**2,590**
	28.58	42.30	26.46	30.82	60.42		**51.78**
Pulmonart valve atresia and stenosis	122	77	14	11	0	218	**442**
	3.81	7.05	3.63	4.84	0.00		**8.84**
Pyloric stenosis	198	24	8	6	0	209	**445**
	6.18	2.20	2.08	2.64	0.00		**8.90**
Rectal and large intestinal atresia / stenosis	74	23	3	4	0	42	**146**
	2.31	2.11	0.78	1.76	0.00		**2.92**
Reduction deformity, lower limbs	24	8	2	0	0	16	**50**
	0.75	0.73	0.52	0.00	0.00		**1.00**
Reduction deformity, upper limbs	38	7	2	0	0	18	**65**
	1.19	0.64	0.52	0.00	0.00		**1.30**
Renal agenesis / hypoplasia	62	15	7	1	1	25	**111**
	1.93	1.37	1.82	0.44	15.11		**2.22**
Spina bifida without anencephalus	60	14	9	2	0	7	**92**
	1.87	1.28	2.33	0.88	0.00		**1.84**
Tetralogy of Fallot	94	27	8	3	0	67	**199**

	2.93	2.47	2.08	1.32	0.00		**3.98**
Transposition of great arteries	67	12	6	0	0	44	**129**
	2.09	1.10	1.56	0.00	0.00		**2.58**
Tricuspid valve atresia and stenosis	11	12	0	3	0	11	**37**
	0.34	1.10	0.00	1.32	0.00		**0.74**
Trisomy 13 (Patau syndrome)	6	6	1	0	0	10	**23**
	0.19	0.55	0.26	0.00	0.00		**0.46**
Trisomy 18 (Edwards syndrome)	19	6	1	0	0	16	**42**
	0.59	0.55	0.26	0.00	0.00		**0.84**
Ventricular septal defect	541	146	55	28	0	543	**1,313**
	16.88	13.37	14.27	12.33	0.00		**26.25**
Total Live Births	320,553	109,219	38,545	22,709	662		**500,234**

Notes:
Amniotic bands data for 2001-2002 only
1998-2002 Total Live Births includes only residents of VA
Data for 2001 & 2002 have not been matched with birth certificates

Appendix C – Figures

Figure A: Data Flow in VISITS / VaCARES



Diagnosis
A physician makes a diagnosis of a birth defect in a child under two during a hospital stay or a genetics center visit.

Report
The ICD9CM code is identified in a discharge report by the hospital VISITS user and entered into the VISITS system.

Analysis
The data in the VISITS / VaCARES system are analyzed at VDH.

Dissemination
VaCARES data is reported annually to NBDPN and to Virginia stakeholders at varied time intervals.

Figure B: Data Quality Analysis – Child Data, 2003-2005

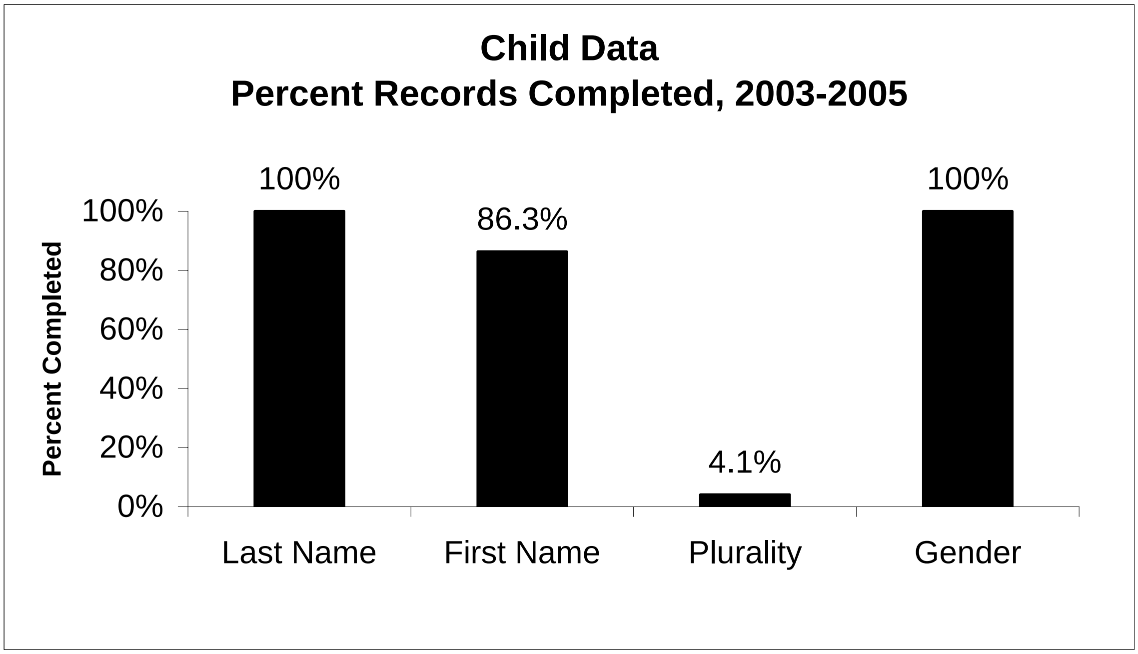


Figure C: Data Quality Analysis – Mother Data, 2003-2005

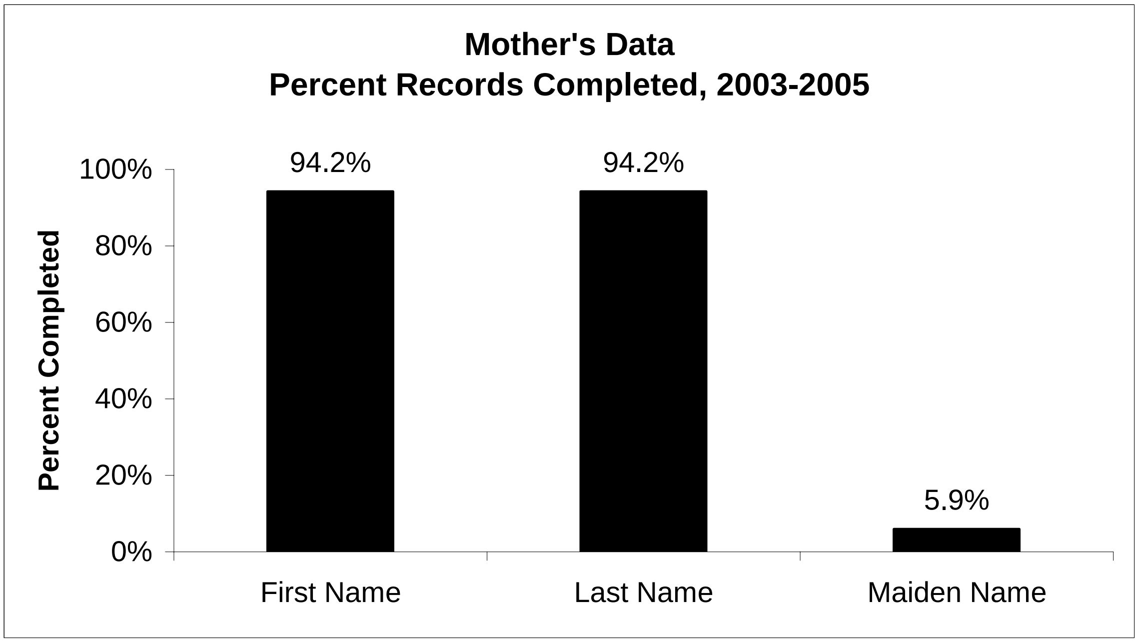


Figure D: Data Quality Analysis – Contact Information, 2003-2005

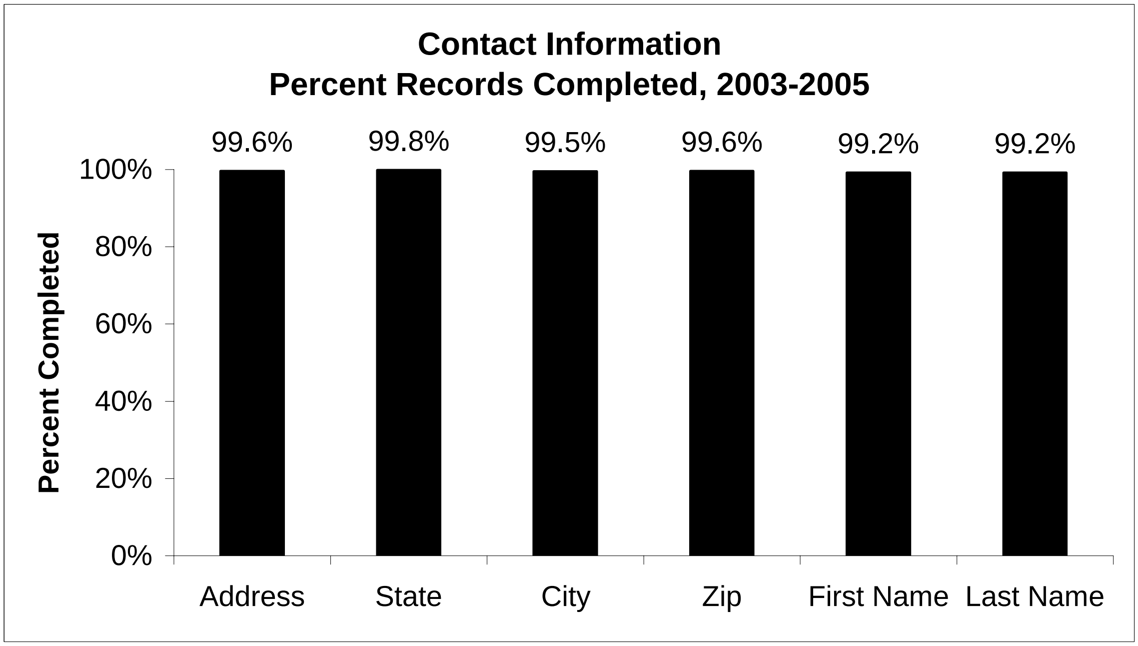


Figure E: Data Quality Analysis – Hospital Information, 2003-2005

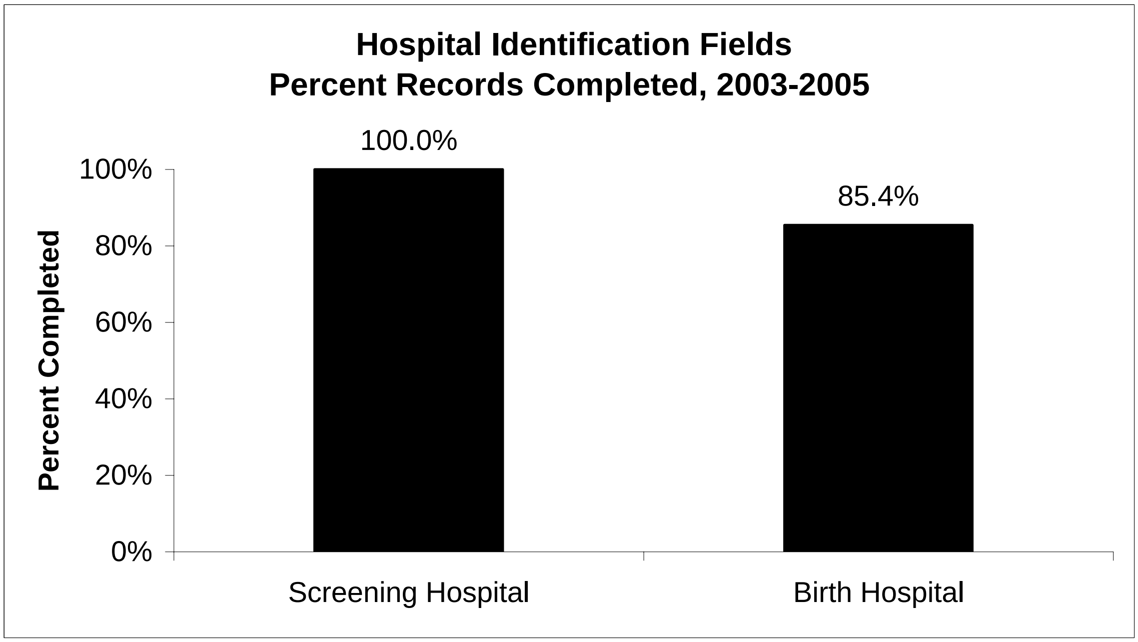


Figure F: Data Quality Analysis – Record Fields, 2003-2005

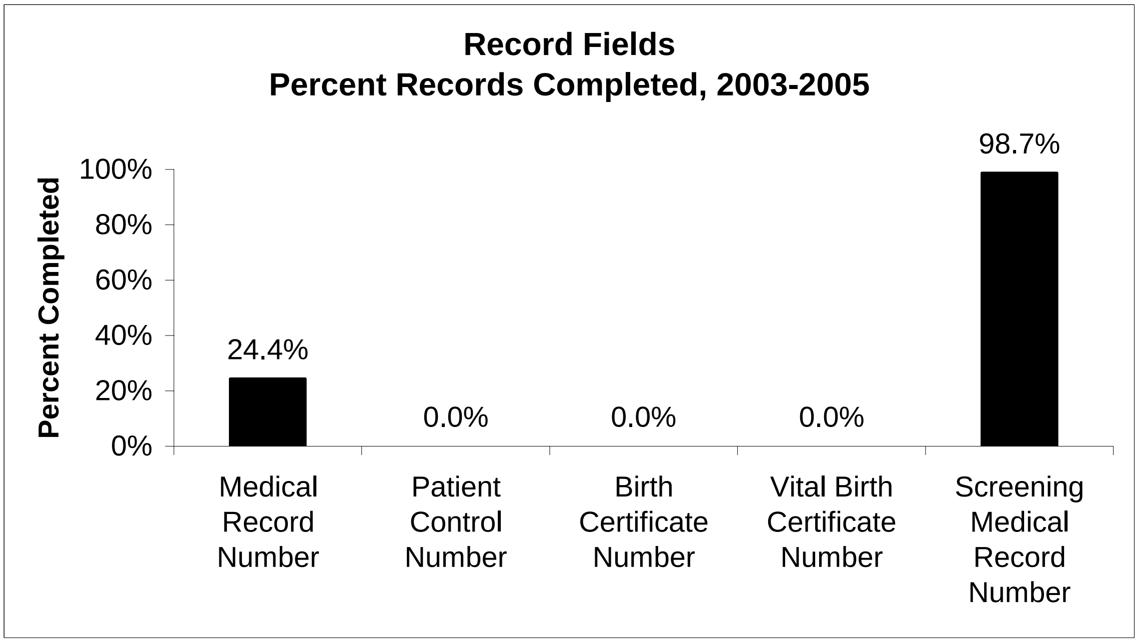


Figure G: Data Quality Analysis – Race / Ethnicity Fields, 2003-2005

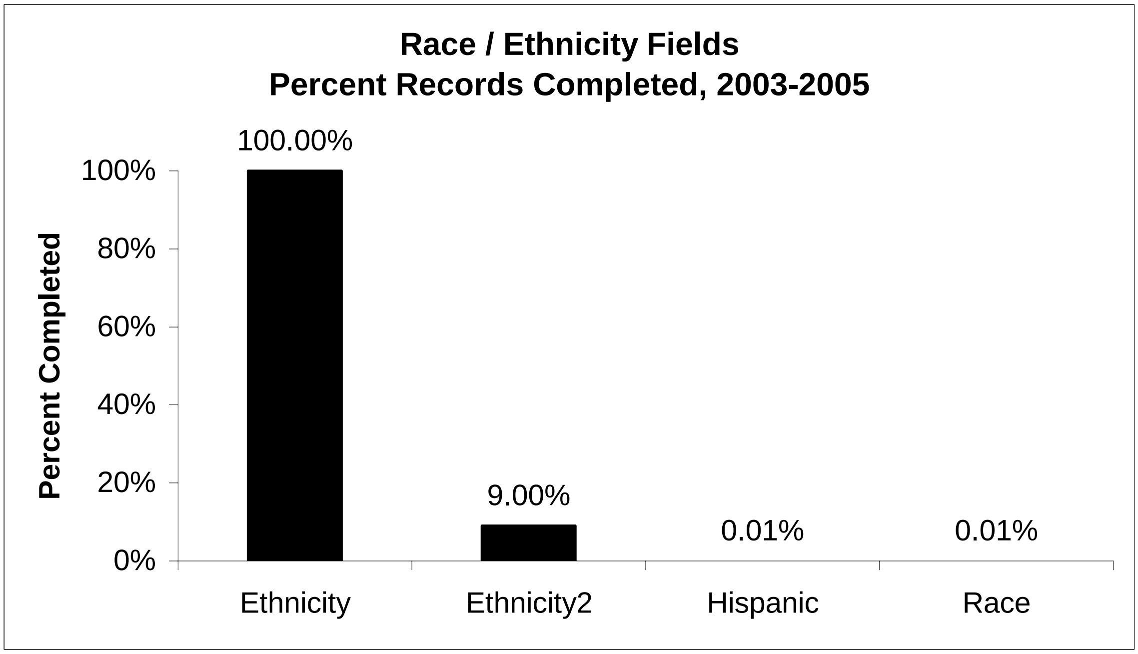


Figure H: Sensitivity (Percent) of VaCARES, 2000-2002

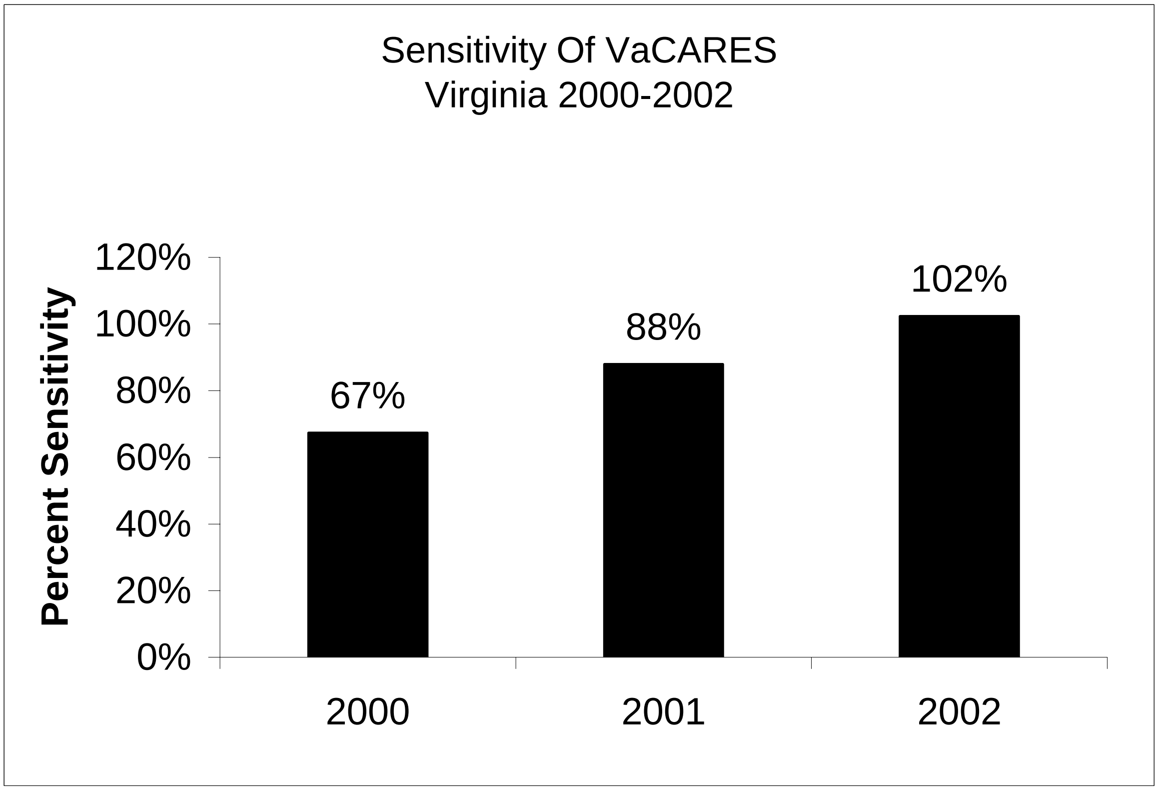


Figure I: Trends in Sensitivity for Five Health Districts Averaging Most Births, 2000-2002

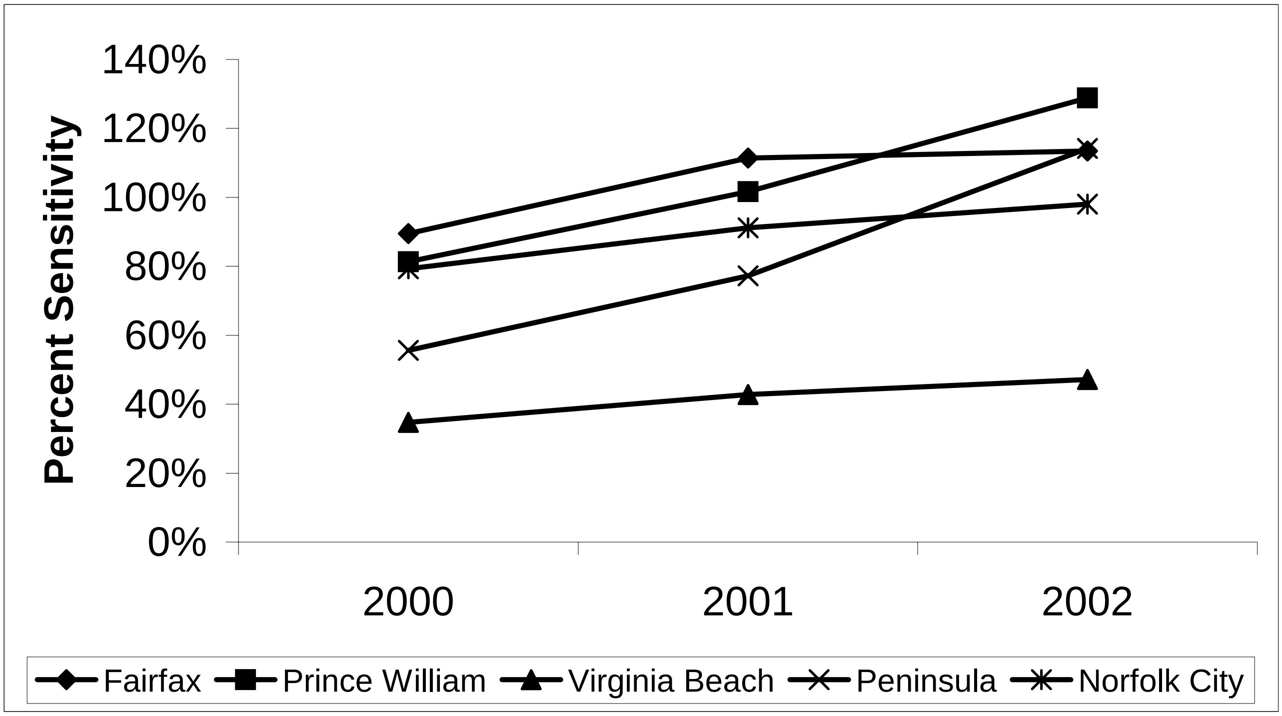


Figure J: Positive Predictive Value, Neural Tube Defects, 2000-2002

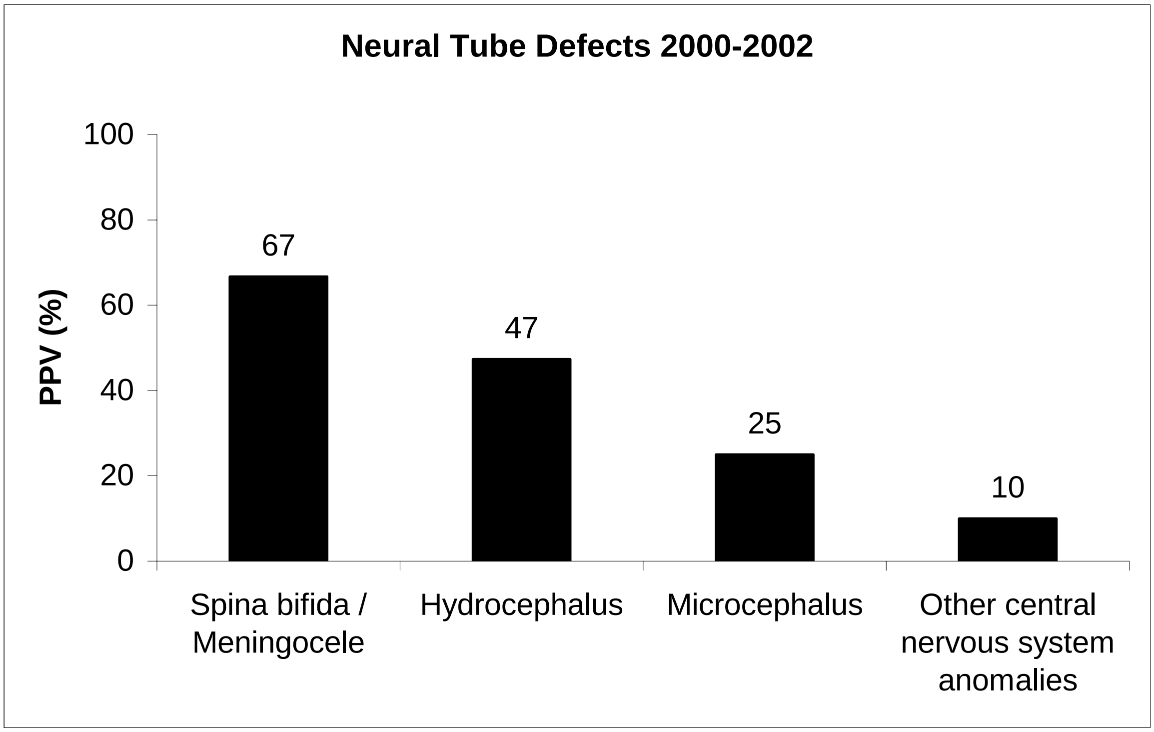


Figure K: Positive Predictive Value, Defects Identifiable at Birth, 2000-2002

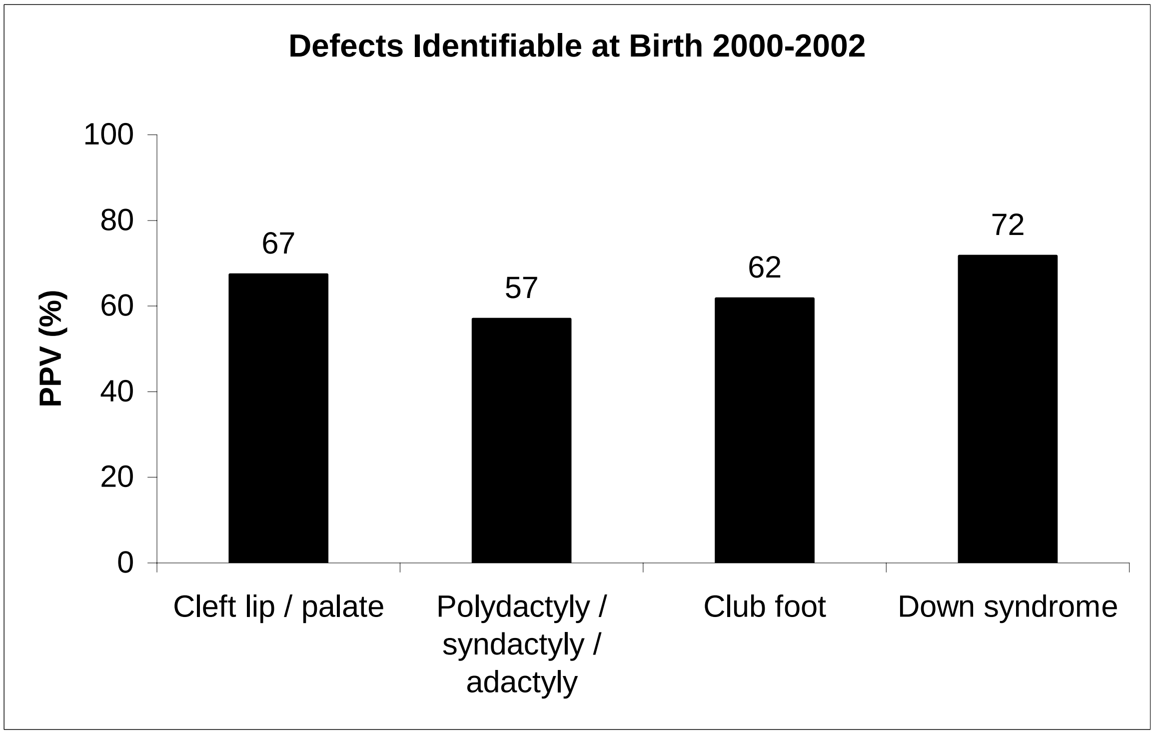


Figure L: Total Number of Children With at Least One Birth Defect, Captured by Birth Certificates and VaCARES, 2000-2003

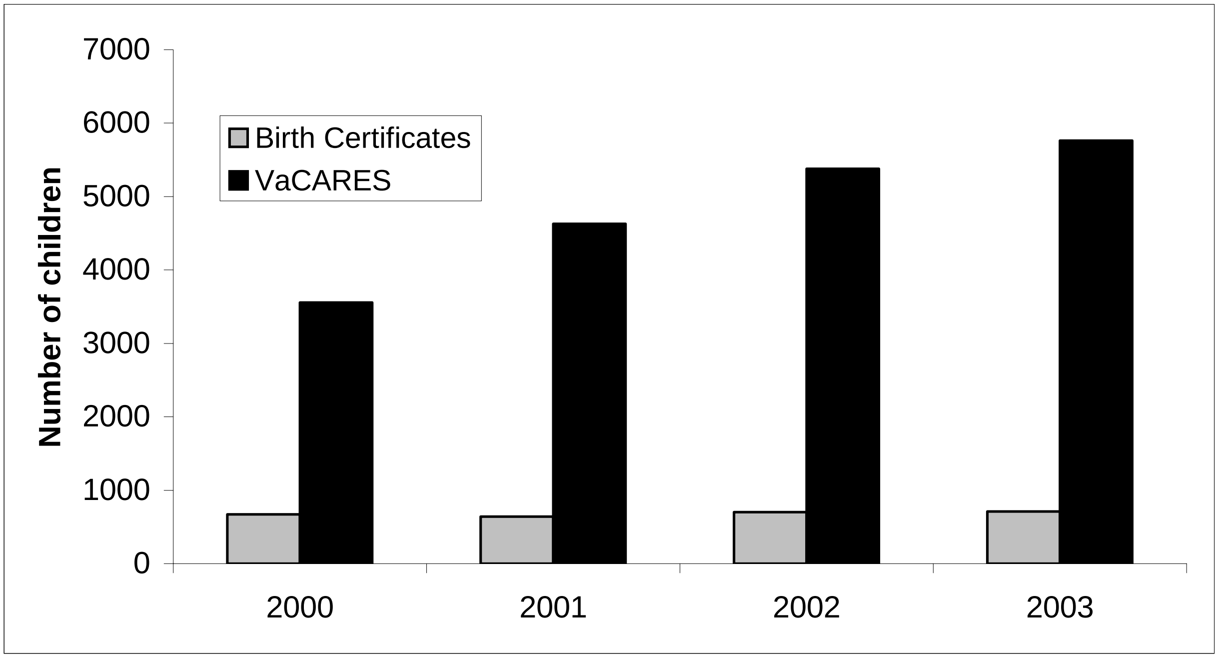


Figure M: Prevalence, Birth Certificates vs. VaCARES 2000-2003

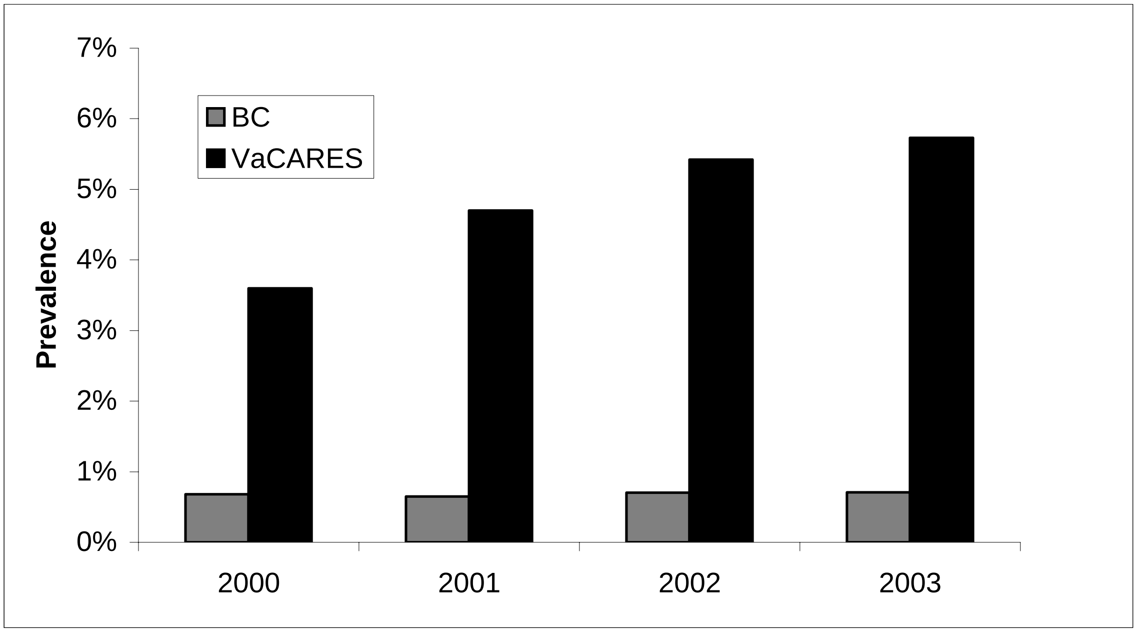


Figure N: Neural Tube Defects, Birth Certificates vs. VaCARES 2000-2003

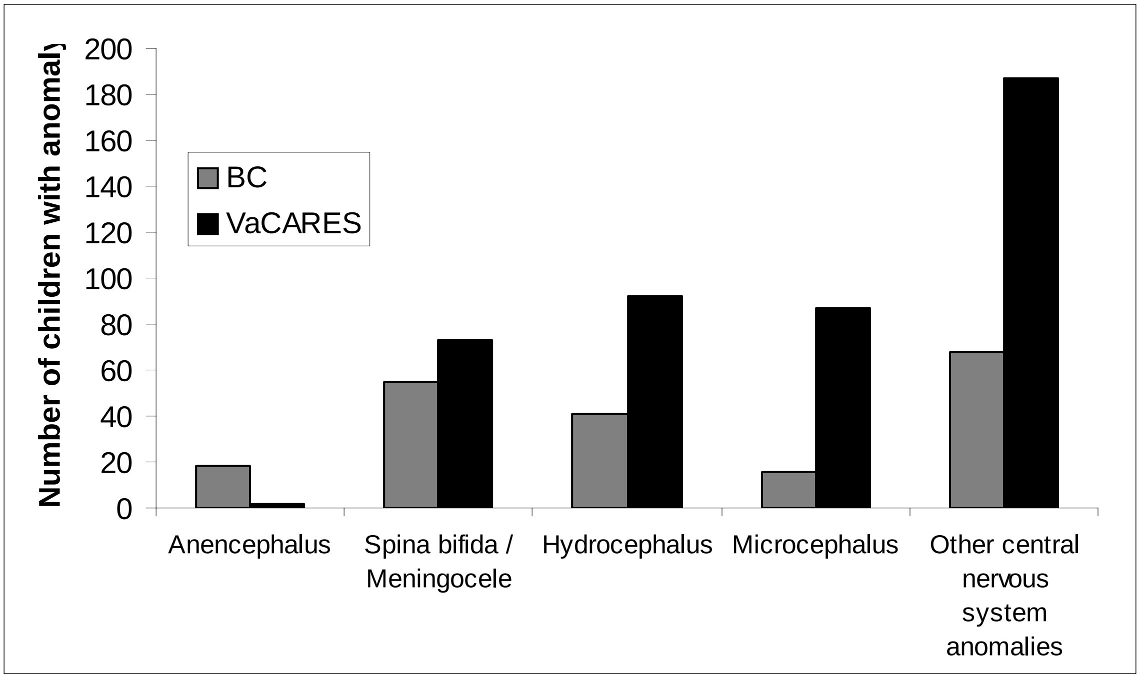


Figure O: Representativeness of VaCARES, All Defects 2000-2002

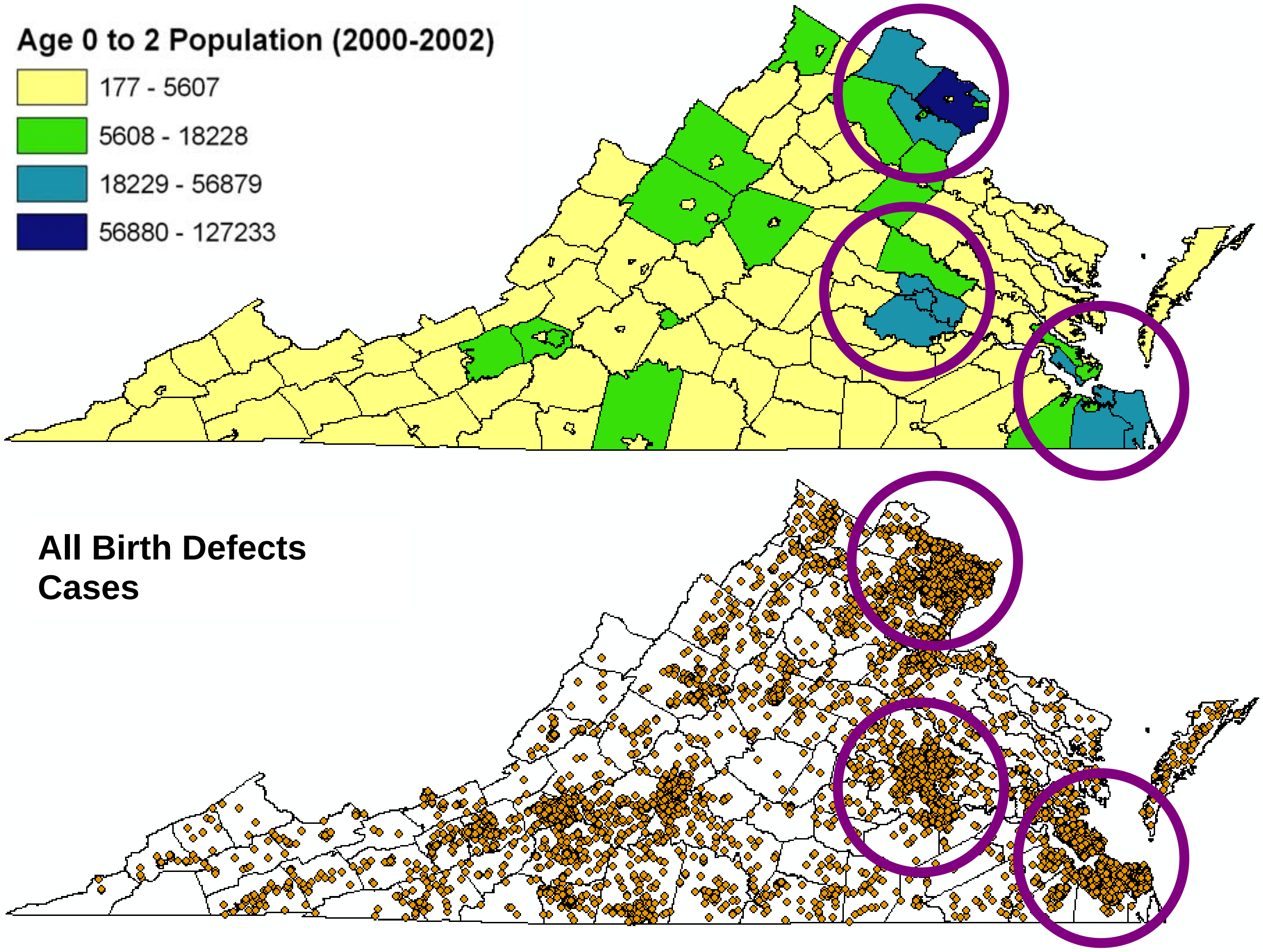


Figure P: Representativeness of VaCARES, Central Nervous System Defects 2000-2002