Congenital Malformations Registry (CMR)

An Update

New York Health Information Management Association

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Director, Congenital Malformations Registry
New York State Department of Health
June 6, 2017
Topics

- CMR Background
- Requirements & Challenges with Birth Defects (BD) Reporting
- Upgrading CMR electronic reporting system
- Describe Birth Defects (BDs) associated with Zika virus
- Recent findings from surveillance of Zika-related defects
  - Microcephaly
  - Other defects
- Recent Zika-related requests for monthly discharge lists and medical records
Why is it important to monitor birth defects?

- Relatively common: 3-5% of live births
- Leading contributor to infant and childhood deaths
- While the majority of children will survive, they and their families may face health, financial and/or other challenges
- Cause of 70% of birth defects is unknown
- Studying birth defects will eventually help us understand what causes them and lead towards prevention
- Hope for prevention → Zika virus
Prozac in Pregnancy May Up Risk of Infant Heart Defects
— Meta-analysis finds significant association with first trimester use of drug

by Molly Walker
Staff Writer, MedPage Today
May 17, 2017

Infants exposed to fluoxetine (Prozac) in their mother’s first trimester of pregnancy had small but significant increased risks of major malformations and cardiovascular malformations, a small systematic review and meta-analysis found.

Economic impact of Zika outbreak could exceed $18B in Latin America, Caribbean

Johns Hopkins researchers conduct rapid assessment analysis based on existing data on incidence, transmission of virus linked to birth defects

https://hub.jhu.edu/2017/05/08/zika-economic-impact-latin-america-caribbean/

https://www.medpagetoday.com/OBGYN/Pregnancy/65380?xid=nl_mpt_DHE_2017-05-18&eun=g1041107d0r&pos=3
Causes of Birth Defects

- Genetic (13-15%)
  - Chromosome number (Down Syndrome, Trisomy 18, Trisomy 13, sex chromosome abnormalities)
  - Chromosome structure (translocations, deletions, duplications)
  - Gene defects (Achondroplasia, Fragile X Syndrome)

- Environmental (7-10%) - the stage at exposure and dosage are very important (Retinoic Acid, Rubella, Thalidomide)

- Multifactorial (20-25%) - threshold model (cleft lip, neural tube defects, pyloric stenosis)

Unknown etiology (50-60%)

Unknown + Multifactorial = (70-85%)
What got Birth Defects Registries started?

- Radiation – 1921
- Rubella – 1940’s
- Thalidomide – late 1950’s

Quarterly thalidomide sales (interrupted line) and numbers of children born with defects characteristic of thalidomide babies (continuous line) ascertained by Lenz (1965) for West Germany excluding Hamburg.

(From Scientific Foundations of Paediatrics, J.A. Davis and J. Dobbing, eds., 1974, p. 749, by permission of William Heinemann Medical Books Ltd.)
Construction of the LaSalle Expressway restricted groundwater from flowing to the Niagara River and the increased precipitation in 1977 led to seepage of canal contents, from degraded drums, to the surface as well as into basements. Residents complain of chemical odors & loss of vegetation.
CMR established under Environmental Disease Surveillance Program in 1982, as a result of “Love Canal” (Niagara Falls, NY)

Migrated to electronic reporting from 2002-2006.

Includes children diagnosed up to 2 years of age, born/residing in NY, with a major birth defect, chromosomal anomaly or persistent metabolic defect. Requires reporting by hospitals & physicians within 10 days of diagnosis.

In NYS, there are about 240,000 births every year. Over 12,000 of these infants will have a major birth defect (about 5% of all births).

May 2016:
• Nurse practitioners, physician assistants, midwives must report
• Reporting of prenatally diagnosed defects required
• Up to age 10 for fetal alcohol syndrome, heart defects, muscular dystrophies and genetic conditions

June 2017:
• Cancers and metabolic defects to be dropped
Goals when conducting surveillance

- Complete
- Accurate
- Timely
- Specific

Large, well-defined population

LOW COST!!
Variation in U.S. State-based BD Surveillance

• Intensive/Active: TX, CA, MA, IA, MI, UT, NC
• Mandatory hospital reporting with follow-up and quality control: NY
• Mandatory hospital reporting without follow-up and quality control
# Birth Defects Rates* determined by various surveillance approaches

<table>
<thead>
<tr>
<th>Method and Source</th>
<th>Rate*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth certificates†</td>
<td>88.9</td>
</tr>
<tr>
<td>Newborn hospital discharge data§</td>
<td>282.5</td>
</tr>
<tr>
<td>Mandatory hospital reporting data•</td>
<td>248.0</td>
</tr>
<tr>
<td>Linked data sources**</td>
<td>336.0</td>
</tr>
<tr>
<td>Active hospital surveillance data‡‡</td>
<td>415.0</td>
</tr>
<tr>
<td>Physical exam of infant§§</td>
<td>830.0</td>
</tr>
</tbody>
</table>

* Per 10,000 births.
† National Center for Health Statistics, CDC, 1982-1983.
• Nebraska Birth Defects Registry, 1982-1985.
BDs where CMR Prevalence Compares favorably to “Active” State Surveillance Programs

<table>
<thead>
<tr>
<th>Birth Defect (Condition)</th>
<th>NYS</th>
<th>Active</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart: Transposition of great arteries</td>
<td>4.5</td>
<td>4.7</td>
<td>4.5-5.0</td>
</tr>
<tr>
<td>Heart: Hypoplastic left heart syndrome</td>
<td>2.4</td>
<td>2.4</td>
<td>2.2-2.6</td>
</tr>
<tr>
<td>Gastrointestinal: Esophageal atresia</td>
<td>2.3</td>
<td>2.4</td>
<td>2.2-2.6</td>
</tr>
<tr>
<td>Chromosomal: Trisomy 21/Down syndrome</td>
<td>12.3</td>
<td>13.7</td>
<td>13.2-14.1</td>
</tr>
</tbody>
</table>

* Prevalence = # of birth defects per 10,000 live births
** CMR birth years 2002-2004
# BDs where CMR Prevalence **DOES NOT** Compare favorably to “Active” State Surveillance Programs

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>NYS</th>
<th>Active</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>CNS: Anencephalus</td>
<td>0.4</td>
<td>2.5</td>
<td>2.3-2.7</td>
</tr>
<tr>
<td>CNS: Spina bifida w/out anencephalus</td>
<td>1.9</td>
<td>3.7</td>
<td>3.4-3.9</td>
</tr>
<tr>
<td>Eye: Anophthalmia/microphthalmia</td>
<td>0.6</td>
<td>2.1</td>
<td>1.9-2.3</td>
</tr>
<tr>
<td>Heart: Endocardial cushion defect</td>
<td>2.9</td>
<td>4.4</td>
<td>4.1-4.6</td>
</tr>
<tr>
<td>Orofacial: Cleft lip w &amp; w/o cleft palate</td>
<td>6.9</td>
<td>10.5</td>
<td>10.1-10.9</td>
</tr>
<tr>
<td>Musculoskeletal: Upper limb reduction</td>
<td>1.8</td>
<td>3.8</td>
<td>3.5-4.0</td>
</tr>
<tr>
<td>Musculoskeletal: Gastroschisis</td>
<td>1.9</td>
<td>3.7</td>
<td>3.5-4.0</td>
</tr>
<tr>
<td>Chromosomal: Trisomy 18</td>
<td>1.1</td>
<td>2.4</td>
<td>2.2-2.6</td>
</tr>
</tbody>
</table>

* Prevalence = # of birth defects per 10,000 live births
** CMR birth years 2002-2004
Diagram illustrating the distortion of the information in a reporting system, from the infant to the coded data. (A) shows the actual infant, (B) is the doctor’s picture of it and what is written down in the medical records, (C) is the content of the report form to the surveillance registry, (D) is the interpretation of that form in the registry, and (E) is the coded data which are stored in the computer.
What happens with reports hospitals submit:

- First review of report:
  - Is BD reportable? At least one major BD? Narrative specific?
  - Match with reports of same infant from other hospitals/prior submissions
- Assign a unique Case Number
- Recode to British Pediatric Association (BPA) system based on narrative
- Processes to improve data quality:
  - Match with vital records (births & deaths)
  - Geocode address of maternal residence
  - Match with SPARCS (hospital discharge data)
  - Hospital discharge summary audit and onsite audits
- Put data to use by summarizing and sharing, and connecting children to Early Intervention
Welcome to the New York State's Congenital Malformations Registry

Birth defects, also called congenital malformations, are a leading cause of infant death in the United States. Every 4.6 minutes, a baby is born with a birth defect.
Choose which type of report you are entering/uploading

To enter cases one at a time, use this option:

Enter Confidential Case Reports

- Routine Current Reports
- Stillbirth Reports
- SPARCS Audit Reports
- Hospital Site Visit Reports
- Hospital Discharge Index
- ECLRS Audit Reports

To upload a file with several cases, use this option:

Upload Case Reports Data

- Routine Current Reports
- SPARCS Audit Reports
- Stillbirth Reports
- Hospital Site Visit Reports
- Hospital Discharge Index
- ECLRS Audit Reports
Report Types

➢ **Routine Current Report** - Almost all of your case reports this type. Choose this if you are sending in everyday, current reports.

➢ **Stillbirth Report** - Choose this if you are submitting a report on a stillborn infant.

➢ **SPARCS Audit Report** – Reports we have asked you to report because a SPARCS Comparison Audit is being conducted. You may see links to the reports on the CMR home page indicating that these cases need to be reported.

➢ **Hospital Site Visit Report** - Following a review of medical records during an in person site visit, unreported/partially-reported cases should be entered here.

➢ **Hospital Discharge Index** - Following a Discharge Summary Audit, a list of unreported cases will be sent back to you and should be reported here.
Enter Routine Current Case Reports

Select the reporting source from the list below: 0001: Albany Medical Center Hospital (Albany)

Enter DOH Date (mm/dd/yyyy): 05/22/2017

Enter Reports
### Help Entering Data

**Routine Current Case Report**

**Child's Information**

<table>
<thead>
<tr>
<th>Field</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>PF1 Number</td>
<td>0001</td>
</tr>
<tr>
<td>Date Reported</td>
<td>05/22/2017</td>
</tr>
<tr>
<td>MRN</td>
<td></td>
</tr>
<tr>
<td>First Name</td>
<td></td>
</tr>
<tr>
<td>Suffix</td>
<td></td>
</tr>
<tr>
<td>AKA/Other Name</td>
<td></td>
</tr>
<tr>
<td>State</td>
<td>NY</td>
</tr>
<tr>
<td>ZIP</td>
<td></td>
</tr>
<tr>
<td>Gestational Age (weeks)</td>
<td></td>
</tr>
<tr>
<td>Birth Status</td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Hispanic</td>
<td></td>
</tr>
<tr>
<td>Birth Order</td>
<td></td>
</tr>
<tr>
<td>Hospital of Birth</td>
<td></td>
</tr>
<tr>
<td>Date of Death</td>
<td></td>
</tr>
<tr>
<td>Foster/Adopted</td>
<td></td>
</tr>
</tbody>
</table>

**Helpful Notes**

- Fill in all mandatory fields (indicated with a star).
- Use the drop-down menus to select values.
- The form is designed to capture comprehensive information about the child's medical history and demographics.
Update Contact Information in CMR Application

HCS and go into the CMR

From the menu on the left, click:

- Update contact information for each role:
  - Director of Health Information Management
  - CMR Registrar
  - SPARCS Outpatient Contact Person
  - Contact Person (for the CMR)
  - Release of Information Officer

Click:

Send Changes to CMR
SPARCS Comparison Audit

- CMR compares reports submitted by your facility with SPARCS inpatient and outpatient data to identify BDs that have not been reported by your facility.
- Upon review of medical records for the child:
  - If child has the BD, the child needs to be reported.
  - If child does not have the BD, please notify us.
  - If child reported previously but specific BD was not reported, we send queries to verify the missing information.

Please review the medical record to verify/respond to queries in a timely manner.

2012 SPARCS Audit Deadline
May 1st, 2017
SPARCS Comparison Audit

- Click on ‘Generate SPARCS Audit Report for 2012.’

- Generated report indicates which records were submitted to SPARCS, but not to the CMR. Click on inpatient and/or outpatient to view outstanding records that need to be reported. We recommend that your facility copy and paste the table of outstanding records into a word or excel document for future reference.

Any outstanding records will be listed in a table.
SPARCS Comparison Audit

- Review outstanding records identified from SPARCS Audit Report. If birth defect is reportable, enter outstanding records as SPARCS Audit Reports under ‘Enter Confidential Case Reports.’

- If birth defect is not reportable, email cmr@health.ny.gov notifying us of the MRN and reason why it is not reportable.

- Please check for and respond to queries as soon as possible. Queries will be sent to your facility if your facility reported information to SPARCS that was not included in the case reports sent to the CMR.
File Upload: Introducing the CSV Option!

-Coming Soon-

➢ Flat file format (files with “.txt” extension) will still be an option but will be removed from the CMR in the future (date TBD)

➢ Please work with your IT team to let them know that the CMR will transition to accepting CSV files only

➢ We will release an updated CMR Handbook with additional information on the new CSV file format requirements
With Coding Manual to be Released in June 2017:

Certain codes to be removed:

- All D codes (diseases of blood/blood-forming organs and disorders involving immune mechanisms) besides D82.1 Di George’s Syndrome
- Most E codes (endocrine, nutritional & metabolic)
- G11 (hereditary ataxias)
- G23-52.7 (many diseases of the nervous system)
- H codes (diseases of the eye)
- P04 (newborn affected by noxious substances transmitted by placenta or breast milk)
To be added to Coding Manual in near future:

Certain codes to be added for fetal diagnosis of BDs from maternal record:

- P95 Stillbirth
- Z37.x Outcome of delivery, stillborn
- O36.4XXx Maternal care for intrauterine death

Some narratives/ICD10 codes have been added to be consistent with the updated 2017 ICD10 Coding Manual and enhanced surveillance for prenatally diagnosed defects.
Challenges with ICD-10 Codes and BD Reporting

<table>
<thead>
<tr>
<th>ICD10</th>
<th>Description</th>
<th>ICD9</th>
</tr>
</thead>
</table>
| Q21.8 | • Other congenital malformations of cardiac septa  
      | • Eisenmenger’s defect  
      | • Pentalogy of Fallot  
      | • Fallot’s tetralogy plus atrial septal defect | 745.8  
      |                                                | 745.2  |
### Other congenital malformations of the brain

<table>
<thead>
<tr>
<th>Q04</th>
<th>Other congenital malformations of the brain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Q04.0</td>
<td>Other anomalies of the corpus callosum</td>
</tr>
<tr>
<td></td>
<td>Agenesis or partial agenesis or absence of corpus callosum</td>
</tr>
<tr>
<td>Q04.3</td>
<td>Other reduction deformities of the brain</td>
</tr>
<tr>
<td></td>
<td>Absence of part of brain</td>
</tr>
<tr>
<td></td>
<td>Agenesis of part brain</td>
</tr>
<tr>
<td></td>
<td>Agyria</td>
</tr>
<tr>
<td></td>
<td>Aplasia of part of brain</td>
</tr>
<tr>
<td></td>
<td>Hydranencephaly</td>
</tr>
<tr>
<td></td>
<td>Hypoplasia of part of brain</td>
</tr>
<tr>
<td></td>
<td>Lissencephaly</td>
</tr>
<tr>
<td></td>
<td>Microgyria</td>
</tr>
<tr>
<td></td>
<td>Pachygyria</td>
</tr>
</tbody>
</table>
### Q04.8 Other specified congenital malformations of the brain

- Arnold-Chiari syndrome, type IV
- Agenesis of part brain
- Agyria
- Aplasia of part of brain
- Hydranencephaly
- Hypoplasia of part of brain
- Lissencephaly
- Microgyria
- Pachygyria
Upgrades to Health Commerce System (HCS)

Effective June 1, 2017, HCS discontinued access for computers running Windows XP and Microsoft Internet Explorer 9 (IE9) and older web browsers as they posed a security risk to HCS applications.
What to do if you experience issues with the CMR application

➢ Email us at cmr@health.ny.gov with detailed description of problem if application is not working as expected. We will work with our IT staff to correct any major issues as quickly as possible.

➢ Common problems outside of the application itself
  ➢ Make sure you’re using most current version of your internet browser
  ➢ Clear your cache
    ➢ For instructions, Google ‘how do I clear my cache for’ and the name of your internet browser (Google Chrome, Mozilla Firefox, Internet Explorer, etc.)
Take home messages

➢ Check for outstanding reports identified by SPARCS Comparison Audit
➢ If you upload files, be sure to sign into CMR and update ‘incomplete’ cases. CMR staff will only process cases that are marked ‘complete.’
➢ Send Reports and Respond to queries thoroughly and timely
➢ Stay current with reporting cases (i.e., close to discharge date)
  ➢ If you have a backlog, please notify us and work with your supervisor/team to find extra help to catch up
➢ Let your IT staff know about new CSV format if you upload files
➢ Email us at cmr@health.ny.gov if you have reporting questions or experience issues with the application
Zika Virus Timeline

- 1947: Zika first discovered in Uganda
- 1970-1980: 14 documented cases of Zika globally in Nigeria, Asia, Indonesia
- 2007: Outbreak in Yap, Pacific Island with 49 confirmed cases; Outbreak in Gabon, Africa
- Ae. albopictus is a vector
- 2013: Outbreak in French Polynesia with 333 confirmed cases; Guillian-Barre Association
- May 2015: Outbreak in Brazil infecting > 200,000
- 2016-2017: Zika identified in many countries, with some newly identified manifestations
Zika in January 2016: we knew very little

- Zika can cause microcephaly
- Zika can cause GBS
- Zika can impact pregnancies at any point in the pregnancy
- Problems can show themselves after birth
- Zika can be sexually transmitted

Zika in June 2017: we know a lot, but still not everything

- How long is someone infectious?
- Does Zika behave differently with coinfections? Cross-reactivity?
- What mosquito species are involved?
- What made Zika so nasty?
- What is the full clinical picture of Zika?
Microcephaly Definition and Diagnosis

• Microcephaly is the clinical finding of a small head compared with infants of the same sex and age, or gestational age, if measured at birth

• Prenatally:
  • Can be difficult to diagnose ★
  • May be detected by routine ultrasound at 18-20 weeks
  • However, best identified on ultrasound later in pregnancy (late second trimester, early third trimester)

• Infant:
  • CDC defines as head circumference < 3rd percentile ★
  • CDC recommends INTERGROWTH-21st ★ standards [http://intergrowth21.ndog.ox.ac.uk/](http://intergrowth21.ndog.ox.ac.uk/)
    • Standard growth charts by gestation, sex, and age

Surveillance is challenging!
Types of microcephaly

- Disproportionate – Head is small out of proportion to the weight and length, which may be normal for age and sex.
- Proportionate – Head size, weight and length are all small for age and sex but proportional to each other.
- “Relative” microcephaly – Head size measures within the normal range for age and sex, but is small out of proportion to the weight and length.

Surveillance is challenging!
Congenital vs. acquired microcephaly

- Congenital microcephaly is usually present prenatally or at the time of birth/delivery
  - Abnormal development of the brain (often genetic)
  - Arrest or destruction of normally-forming brain (e.g., infection, vascular disruption)
- Acquired microcephaly develops as a result of late onset infection/insult (e.g., perinatal stroke), or after birth due to delivery complications or postnatal insult, trauma or infection
  - HC is normal at birth
  - As the baby grows in length, the head becomes comparatively smaller

Surveillance is challenging!
Causes of Congenital Microcephaly

– Genetic causes
  • Single gene disorders (syndromes)
  • Chromosomal abnormalities, microdeletions, microduplications
  • Mitochondrial mutations
– *In utero* ischemia/hypoxia (e.g., placental insufficiency or abruption)
– Teratogens (e.g., maternal alcohol, hydantoin)
– Radiation
– Mercury (e.g., fish and seafood)
– Maternal conditions (e.g., poorly controlled diabetes, hyperphenylalaninemia)
Causes of Congenital Microcephaly

– *In utero* infections
  • Toxoplasmosis
  • Rubella
  • Cytomegalovirus (CMV)
  • Herpes
  • Human Immunodeficiency Virus (HIV)
  • Syphilis
  • Zika virus 4/2016!

Fetal brain disruption sequence (FBDS) – No ICD10 code yet !!
Study of Zika virus infection in Pregnant women in Brazil (Sept. 2015-May 2016)

Features differentiating Congenital Zika Virus Syndrome (CZVS) from other congenital infections:

- Severe microcephaly with partially collapsed skull
- Thin cerebral cortices with subcortical calcifications
- Macular scarring and retinal pigment epithelial abnormalities
- Congenital contractures
- Marked early hypertonia with symptoms of extrapyramidal involvement

Congenital Zika Virus Syndrome

Destruction of existing CNS tissue and Disruption of future developmental processes

Loss of brain volume

Neurologic dysfunction

Severe microcephaly
- Misshapen skull with overlapping sutures
- Redundant scalp

Hearing, vision, swallowing problems
- Global developmental impairment
- Limb contractures
- Hypertonia, epilepsy, extreme irritability

Recognizable pattern = Congenital Zika Syndrome

Courtesy of CDC: https://www.youtube.com/watch?v=6P6008JbfIE
Cranial Morphology

- Partial collapse of the skull with overlapping sutures
- Consistent with fetal brain disruption sequence (FBDS)
- Not all severe microcephaly will look like this

Courtesy of CDC: https://www.youtube.com/watch?v=6P6008JbfIE
Zika-Related Brain Abnormalities with/without Microcephaly

- Intracranial calcifications
- Cortical gyral abnormality
- Porencephaly
- Hydranencephaly
- Ventriculomegaly
- Intraventricular Hemorrhage
- Fetal Brain Disruption Sequence
Eye Abnormalities

- Microphthalmia
- Coloboma
- Chorioretinal atrophy
- Optic Nerve Atrophy
- Congenital Cataract
- Intraocular calcifications
Birth Defects Among Fetuses/Infants of US Women With Evidence of Possible Zika Virus Infection During Pregnancy

• Among pregnant women in U.S. with completed pregnancies and lab evidence of possible Zika infection, 6% of fetuses or infants had evidence of Zika-associated birth defects, primarily brain abnormalities & microcephaly.

• Of women with first-trimester Zika infection, 11% of fetuses or infants had evidence of Zika-associated birth defects.

## Impact of Zika on New Yorkers

### US States

<table>
<thead>
<tr>
<th>Category</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Locally acquired cases reported</td>
<td>224</td>
</tr>
<tr>
<td>Travel-associated cases reported</td>
<td>5,013</td>
</tr>
<tr>
<td>Acquired through other routes</td>
<td>48</td>
</tr>
</tbody>
</table>

**Total Cases as of May 31, 2017**

<table>
<thead>
<tr>
<th>Category</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pregnant women</td>
<td>1,883</td>
</tr>
<tr>
<td>Completed pregnancies</td>
<td>1,579</td>
</tr>
<tr>
<td>Liveborn with birth defects</td>
<td>72</td>
</tr>
<tr>
<td>Pregnancy losses with birth defects</td>
<td>8</td>
</tr>
</tbody>
</table>

### NYS

<table>
<thead>
<tr>
<th>Category</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Locally acquired cases reported</td>
<td>0</td>
</tr>
<tr>
<td>Travel-associated cases reported</td>
<td>1,397</td>
</tr>
<tr>
<td>NYC</td>
<td>1,076</td>
</tr>
<tr>
<td>Outside NYC</td>
<td>321</td>
</tr>
<tr>
<td>(28%)</td>
<td></td>
</tr>
</tbody>
</table>

| Acquired through other routes      | 5      |

**Total Cases as of June 2, 2017**

<table>
<thead>
<tr>
<th>Category</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pregnant women</td>
<td>498</td>
</tr>
<tr>
<td>NYC</td>
<td>402</td>
</tr>
<tr>
<td>Outside NYC</td>
<td>96</td>
</tr>
<tr>
<td>(26%)</td>
<td></td>
</tr>
</tbody>
</table>

| Completed pregnancies             | -      |
| Liveborn with birth defects       | 8      |
| Pregnancy losses with birth defects| -      |
CMR Response to Zika

• Established baseline prevalence for microcephaly (births 2013-15)
  – Required clear microcephaly classification
  – Medical record review was a necessity

• Active surveillance of defects in newborns with medical record review

• Active surveillance of prenatally diagnosed defects (with maternal medical record review)

• Connecting families of affected newborns to Early Intervention services
“Newborn” Microcephaly Cases by Birth Year, from SPARCS Inpatient (1990-2014)

150 cases

CRUDE Microcephaly prevalence: 2-12/10,000 live births (US) (for births in 2009-13) 4.6-6.7/10,000 live births (NY)

**NYS Live Births:**
- 2013: 236,980
- 2014: 238,773
- 2015: 233,873
- Total: 709,626

**Total Identified by Hospitals**
(N=489)

**Crude Microcephaly**
6.6/10,000 live births
(n=465)

**Exclusions:**
- Acquired Microcephaly (n=6)
- Possible Microcephaly (n=5)

**Severe Microcephaly, HC <3%**
3.6/10,000 live births
(n=259; 57%)

**Less Severe Microcephaly, 3% ≤ HC ≤ 5%**
0.7/10,000 live births
(n=49; 11%)

**Other Microcephaly, HC >5%**
- Median: 12%
- Range: 5.1% – 99%
2.1/10,000 live births
(n=146; 32%)

**No Documented Physician Diagnosis and No Abstractor Diagnosis**
(n=24)

**NYS Live Births:**
- 2013: 236,980
- 2014: 238,773
- 2015: 233,873
- Total: 709,626

<table>
<thead>
<tr>
<th>Location</th>
<th>2013</th>
<th>2014</th>
<th>2015</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>New York State</td>
<td>81(3.4)</td>
<td>79(3.3)</td>
<td>99(4.2)</td>
<td>259(3.6)</td>
</tr>
<tr>
<td>New York City</td>
<td>52(4.7)</td>
<td>50(4.5)</td>
<td>55(5.0)</td>
<td>157(4.7)</td>
</tr>
<tr>
<td>Health Service Area</td>
<td># of newborns with microcephaly</td>
<td># of Births</td>
<td>Prevalence (per 10,000 live births)</td>
<td></td>
</tr>
<tr>
<td>---------------------------</td>
<td>---------------------------------</td>
<td>-------------</td>
<td>------------------------------------</td>
<td></td>
</tr>
<tr>
<td>Western NY</td>
<td>33</td>
<td>45,914</td>
<td>7.2</td>
<td></td>
</tr>
<tr>
<td>Finger Lakes</td>
<td>23</td>
<td>39,301</td>
<td>5.9</td>
<td></td>
</tr>
<tr>
<td>Central NY</td>
<td>16</td>
<td>45,412</td>
<td>3.5</td>
<td></td>
</tr>
<tr>
<td>NY-Penn</td>
<td>2</td>
<td>8,547</td>
<td>2.3</td>
<td></td>
</tr>
<tr>
<td>Northeastern NY</td>
<td>7</td>
<td>40,676</td>
<td>1.7</td>
<td></td>
</tr>
<tr>
<td>Mid-Hudson</td>
<td>22</td>
<td>70,512</td>
<td>3.1</td>
<td></td>
</tr>
<tr>
<td>NYC</td>
<td>162</td>
<td>336,047</td>
<td>4.8</td>
<td></td>
</tr>
<tr>
<td>Nassau-Suffolk</td>
<td>19</td>
<td>86,668</td>
<td>2.2</td>
<td></td>
</tr>
<tr>
<td><strong>ALL AREAS</strong></td>
<td><strong>284</strong></td>
<td><strong>673,077</strong></td>
<td><strong>4.2</strong></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Census Tract Poverty Level</th>
<th>Prevalence (per 10,000 live births)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very High Poverty</td>
<td>7.2</td>
</tr>
<tr>
<td>High Poverty</td>
<td>4.5</td>
</tr>
<tr>
<td>Medium Poverty</td>
<td>3.7</td>
</tr>
<tr>
<td>Low Poverty</td>
<td>2.7</td>
</tr>
<tr>
<td>NYC</td>
<td>4.7</td>
</tr>
</tbody>
</table>
Current Zika-study Goals: Determine prevalence of birth outcomes and birth defects related to Zika from Jan. 2016 onward

“Active surveillance”

CDC provided a list of Birth Defects Potentially Linked to Zika Virus (Case Inclusion Guidance for Medical Record Abstraction)

Requests for monthly discharge summaries and medical records
Surveillance for Prenatal Zika-related Defects Challenging

O35.xXXx Maternal care for known/suspected fetal abnormality
  • O35.0XXx CNS malformation (7%)
  • O35.1XXx Chromosomal malformation (2%)
  • O35.3XXx Fetal damage from maternal viral disease (CMV, Rubella)
  • O35.8XXx Other fetal damage from listeriosis & toxoplasmosis (78%)
  • O35.9XXx Fetal anomaly, damage unspecified (12%)

O36.4XXx Maternal care for intrauterine death, fetal death NOS, fetal death after 20 weeks gestation, late fetal death, or missed delivery

Z37.x Stillbirths

2016 SPARCS Inpatient Discharges for O35.xXXx and Z37.x: 1,354
### Report of Zika-related BDs, 2013-14*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Brain abnormality or microcephaly</th>
<th>NTDs &amp; other early brain malfs.</th>
<th>Eye abnormalities</th>
<th>Other CNS dysfunction</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td># infants/fetuses (n=747)</td>
<td>392 (52%)</td>
<td>229 (31%)</td>
<td>81 (11%)</td>
<td>45 (6%)</td>
<td>100%</td>
</tr>
<tr>
<td>Pregnancy outcome:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Live birth</td>
<td>89%</td>
<td>52%</td>
<td>100%</td>
<td>96%</td>
<td>79%</td>
</tr>
<tr>
<td>Loss</td>
<td>11%</td>
<td>48%</td>
<td>0%</td>
<td>4%</td>
<td>21%</td>
</tr>
<tr>
<td>Earliest age defect noted (n=410):</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prenatally</td>
<td>55%</td>
<td>89%</td>
<td>7%</td>
<td>18%</td>
<td>56%</td>
</tr>
<tr>
<td>&lt;=28 days of delivery</td>
<td>27%</td>
<td>8%</td>
<td>54%</td>
<td>70%</td>
<td>28%</td>
</tr>
<tr>
<td>&gt;28 days of delivery</td>
<td>18%</td>
<td>4%</td>
<td>39%</td>
<td>11%</td>
<td>17%</td>
</tr>
<tr>
<td>Prevalence/1,000 live births</td>
<td>1.50</td>
<td>0.88</td>
<td>0.31</td>
<td>0.17</td>
<td></td>
</tr>
<tr>
<td>Expected #s in NYS Pre-Zika</td>
<td>360</td>
<td>211</td>
<td>74</td>
<td>41</td>
<td>686</td>
</tr>
</tbody>
</table>

CMR Connecting Families to Early Intervention (EI) Services

- Match newborns with Zika-related birth defects to EI
- If no referral made, EI Program in Albany will contact EI staff in local health departments to request follow up
Discussion Topics

• Challenges with:
  – monthly reporting of Zika-related birth defects with medical records access?
  – reporting prenatally diagnosed defects through maternal record?

• Suggestions for improving the CMR Application?
Additional Information about Zika virus

New York State Department of Health:

Centers for Disease Control and Prevention
Thank you!

QUESTIONS??

Contact us at cmr@health.ny.gov