Thursday 28th April – Morning

Poster Screen 1:

74 Congenital Diaphragmatic Hernia: What can the National Congenital Anomaly and Rare Disease Registration Service (NCARDRS) tell us?

Dr Kathryn Johnson[2], Jennifer Brougham[2], Corinne Mallinson[2], Steven Hardy[2], Dr Kathryn Johnson
[1]Leeds Teaching Hospitals Nhs Trust, [2]National Congenital and Rare Disease Registration Service

110 Guideline revision and care standardization results in improved survival and decreased ECMO utilization for congenital diaphragmatic hernia (CDH) patients

Patrick Sloan[1], NNP-BC Kristen Bagby[2], Dr. Nathalie El Ters[1], Dr. Tasnim Najaf[1]
[1]Washington University in St. Louis, Department of Pediatrics, Division of Newborn Medicine, [2]Saint Louis Children's Hospital

74

Congenital Diaphragmatic Hernia: What can the National Congenital Anomaly and Rare Disease Registration Service (NCARDRS) tell us?

Dr Kathryn Johnson[2], Jennifer Brougham[2], Corinne Mallinson[2], Steven Hardy[2], Dr Kathryn Johnson
[1]Leeds Teaching Hospitals Nhs Trust, [2]National Congenital and Rare Disease Registration Service

BACKGROUND
NCARDRS is a registration service that collects and quality assures data on congenital anomalies and rare diseases in England. NCARDRS forms part of the National Disease Registration Service (NDRS); previously established by Public Health England and now maintained by NHS Digital.
Antenatal screening for Congenital Diaphragmatic Hernia (CDH) forms part of the NHS Fetal Anomaly Screening programme (FASP). As NCARDRS collects information on anomalies collected both antenatally and postnatally, NCARDRS works closely with the NHS Fetal Anomaly Screening Programme (FASP) to audit the detection of the 15 conditions included in the 20 week scan, including CDH.

We aim to describe the live birth prevalence of CDH from 2015 – 2019 in England along with genetic data and data for associated anomalies.

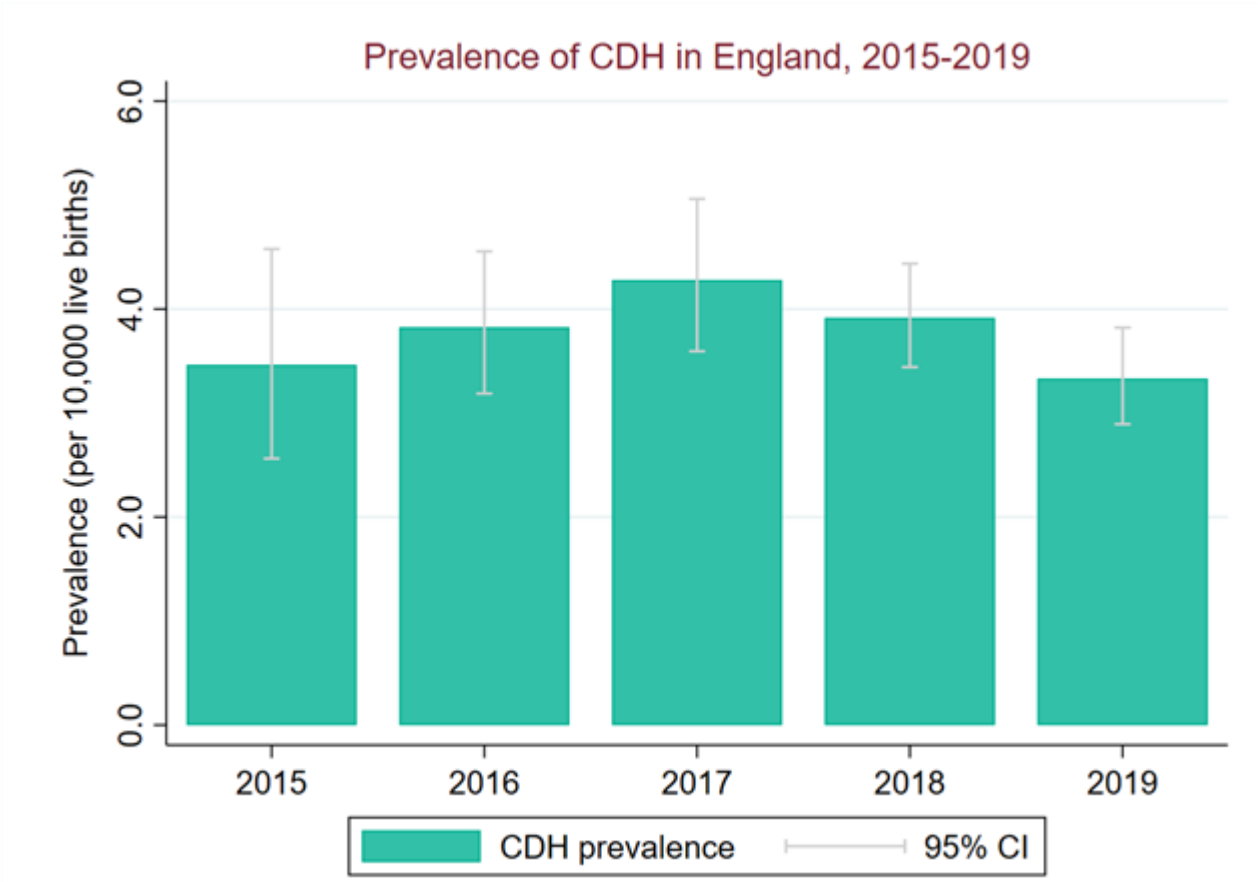
METHODS
Using NCADRS data the prevalence of CDH in babies born 2015-2019 in reporting regions of NCARDRS was calculated per 10,000 live and still births.

RESULTS
See image
The commonest associated congenital anomaly was a cardiac defect with 22.4% of cases (live, still born, terminations or miscarriages >24 weeks gestation) having associated CHD.
Of those who had a genetic test, 20% of the cases in the dataset, had an abnormal test.

DISCUSSION
Our data show the current live birth prevalence of CDH in England; NCARDRS can link this data to other routinely collected data building a clinical picture of CDH after birth and in to childhood, in addition to basic prevalence data.

CONCLUSION
The live birth prevalence of CDH in England is 3-4/10,000.
CDH is not infrequently associated with chromosomal abnormalities and other congenital anomalies.
NCADRS ability to provide national prevalence data on the condition along with linkage to multiple other data sources provides an opportunity to obtain valuable information on the potential causes of congenital anomaly, clinical treatment, associated morbidities and outcomes into adulthood for these children.

Images


Prevalence of CDH in England, 2015-2019
Prevalence (per 10,000 live births)
0.0
2.0
4.0
6.0
2015
2016
2017
2018
2019
CDH prevalence
95% CI

110

Guideline revision and care standardization results in improved survival and decreased ECMO utilization for congenital diaphragmatic hernia (CDH) patients

Patrick Sloan[1], NNP-BC Kristen Bagby[2], Dr. Nathalie El Ters[1], Dr. Tasnim Najaf[1]
[1]Washington University in St. Louis, Department of Pediatrics, Division of Newborn Medicine, [2]Saint Louis Children's Hospital

Background: In 2020 we revised our CDH guidelines to emphasize early treatment of cardiac dysfunction with inotropic support, volume ventilation for a gentle ventilation strategy, and a dedicated care team of neonatologists, nurse practitioners, nurses, and respiratory therapists to standardize and improve care.

Materials and Methods: A retrospective chart review from 2015 – 2021 was undertaken at a Level IV NICU. All CDH infants with active resuscitation are included in this analysis. A total of 66 patients are included; 16 patients with the new guidelines and 50 patients in the historical group. Statistical analysis performed with Fisher's exact test and t-test.

Results: Prenatal indices and liver herniation were similar between the two groups. Cardiac dysfunction was present in 56% (9) of the post guideline group and 40% (20) of the historical group (p=0.38). Survival in the post guideline group was 88% (14) compared to 64% (32) of the historical cohort (p=0.11). ECMO utilization in the post guideline group was 31% (5) and was 58% (29) in the historical group (p=0.08). ECMO survival in the post-guideline group was 80% (4) compared to 45% (13) in the historical group (p=0.33). The average number of days on ECMO was 9 in the post-guideline group compared to 22 in the historical group (p=0.07).

Conclusion: In this small single-center cohort we demonstrate trend towards improved survival and decreased ECMO utilization. We standardized care with a dedicated team delivering care from the attending to the bedside nurse. The improved trend in survival is likely due to a decrease in ECMO utilization, decreased days on ECMO as well as improved survival for the patients requiring ECMO. Overall it is unlikely that one single factor can explain our improved outcomes, rather it is the comprehensive team approach focusing on cardiac support, lung-protective ventilation, and pulmonary hypertension management.