CRANE Database
T: 020 7869 6610
E: crane@rcseng.ac.uk
W: www.crane-database.org.uk

CLEFT REGISTRY & AUDIT NETWORK

CRANE Database: Syndromes and additional diagnoses

Prepared by the CRANE Database Project Team

July 2021

From January 2021 the CRANE Database changed the way it captured data on syndromes and additional diagnoses.

This document, for cleft team members involved in CRANE data collection/data entry, (a) describes this update to the CRANE Database, and (b) provides guidance on how information on syndromes and additional diagnoses is now collected.

Prior to January 2021, the **Syndromes** section of the CRANE database used to require cleft teams to enter free text to describe named syndromes and descriptions of affected systems. This information is now captured by clear categories. The questions and potential responses are set out below.

Confirmed syndrome or significant additional diagnosis present?
>   1=No
>   2=Yes, named
>   3=Yes, unknown name

a) Main syndrome or additional diagnosis: The 9 most common syndromes/significant additional diagnoses affecting children with a cleft were identified from the free text entries on the CRANE database, as well as from a CRANE dataset linked to Hospital Episode Statistics records. If the child's diagnosis does not fit into one of the following 9 categories, then the 10th option 'other' can be selected and free text can be entered.

>   *Main syndrome or additional diagnosis*
>   1 = CHARGE syndrome
>   2 = Chromosome or gene abnormalities not elsewhere specified (e.g. trisomy, deletion, duplication)
>   3 = Congenital malformations of the circulatory system (arteries, veins or heart)
>   4 = Congenital malformations of the nervous system (e.g. microcephaly, spina bifida)
>   5 = Di George syndrome/22q11.2 Deletion syndrome / Velocardiofacial syndrome
>   6 = Fetal alcohol syndrome
>   7 = Goldenhar syndrome / Hemi-facial microsomia
>   8 = Stickler syndrome
>   9 = Van der Woude syndrome
>   10 = Other– please specify: _____ (free text)

b) Secondary syndrome or additional diagnosis: In the unlikely event that a child has been diagnosed with two syndromes or significant additional diagnoses, the list above is repeated under the heading *Secondary syndrome or additional diagnosis.* Cleft teams can then select an option or choose 'other' and enter free text.

Please do not enter typical features of a syndrome in addition to the main syndrome. For example, if a child has been diagnosed with CHARGE syndrome and this is entered as the 'main syndrome or additional diagnosis', please do not enter congenital malformations of the circulatory system as a 'secondary syndrome or additional diagnosis'.

c) Specify affected system(s): Instead of a free text box to collect 'Description of affected systems', there is now a field that states *If syndrome is 'Yes, unknown name' please specify affected system(s):* (Tick all that apply)

       1 = Circulatory/cardiovascular system
       2 = Digestive/excretory system
       3 = Endocrine system
       4 = Nervous system
       5 = Renal system
       6 = Respiratory system
       7 = Skeletal system
       8 = Other– please specify: _____ (free text)

Important guidance on recording syndromes

Pierre Robin Sequence - PRS: Please do not record Pierre Robin Sequence (or any PRS features) as a syndrome or significant additional diagnosis. PRS information should be entered under the ***Cleft Details*** tab.

DIAGNOSES NOT TO BE ENTERED UNDER THE *SYNDROMES* SECTION: Developmental delay and autism should not be entered into the information captured under the ***Syndromes*** tab. Minor issues, such as tongue tie or skin tags should not be entered either.

Notes

MIGRATION OF DATA IN 2020: At the end of 2020, CRANE reviewed all CRANE records with syndrome information entered. Each record had this information re-coded according to the new categories. Any cases whose syndrome/additional diagnoses did not fit into one of the first nine categories was re-coded as '10 – other' and the free text entry was migrated to the new system.

FUTURE ANALYSES AFTER 2021: We will liaise with relevant Clinical Excellence Networks (CENS), including Speech and Language Therapy, to determine which syndromes/additional diagnoses from the list should potentially be included/excluded when reporting each cleft-related outcome. This update to the database will provide much more flexibility when performing analyses.

Thank you very much for your work with the CRANE Database project.

If you have any questions about this User Guide, or wish to provide us with any feedback on this document, please contact us on 020 7869 6610 or crane@rcseng.ac.uk.