Eligibility for the CHAANZ CHD Registry

## 1.1 Diagnosis Reference Table

The CHAANZ Diagnosis Reference Table (DRT), which is centred on EPCC codes, is submitted separately in electronic format and comprises the following variables:

* **CHAANZ\_source:** Tracks changes to the DRT eligibility rules through different versions, where ‘Drt\_sl\_24aug2021’ is the first version, and related files are archived here: Heart Research Institute\Clinical Research Group - CHAANZ\Group A\Diagnosis Coding\02\_DRT\Archive.
* **DIAGNOSIS\_R:** The EPCC name of the diagnosis.
* **IPCCC\_CODE\_R:** The EPCC code for the diagnosis. Note that ‘IPCCC’ refers to an older version of the registry where the IPCCC long list was used. The current DRT, CHAANZ Registry and the Procedure Reference Table described below now all use the EPCC list.
* **CHAANZELIGIBILITY:** Binary flag to denote baseline eligibility for the registry. If a patient has at least one diagnosis where CHAANZELIGIBILITY = 1, they are eligible. The only exception is described in Section 2.1 Paediatric Rules, where diagnosis codes accompanied by the rules ‘PFO’, ‘PDA’ or ‘VALVE’ denote an additional step applied to patients from child hospitals. For these diagnoses, patient participation in the Registry is revoked if no other diagnoses where CHAANZELIGIBILITY = 1 are present. This exception is described in detail below.
* **GENE\_FLAG:** Binary flag for diagnoses to indicate that the diagnosis is a genetic condition. GENE\_FLAG applies to individual diagnosis codes and is used to generate two patient level flags: Gene\_Patient and Gene\_Isolated. If a patient has one or more diagnoses where GENE\_FLAG = 1, their Gene\_Patient flag equals 1 regardless of all other diagnoses. In addition, a patient that only has eligible diagnoses that have GENE\_FLAG = 1, they are flagged separately as Gene\_Isolated = 1. This indicates that the only diagnosis that qualifies them for the Registry is a genetic diagnosis.
* **RULE:** When RULE = 1, the diagnosis is accompanied by a special rule that calculates patient eligibility in the context of all their other diagnoses.
* **RULE\_TYPE:** Indicates the type of rule that accompanies the diagnosis and is only filled in when RULE = 1. When RULE = 1, RULE\_TYPE is either GENE\_SPECIFIC, PFO, PDA, or VALVE.
* **ASSOCIATED\_CODES:** ASSOCIATED\_CODESis a special variable that applies when RULE\_TYPE = GENE\_SPECIFIC. This only occurs for three codes: Marfan’s, Williams and Loeys\_Dietz syndromes. When a patient has one or more of these syndromes, we consider their other diagnoses and look for codes listed in the ASSOCIATED\_CODES variable. This ASSOCIATED\_CODES variable lists codes that must be present for a patient with the for the genetic condition to be eligible for inclusion in the Registry (in the absence of baseline eligibility).
* **sts\_code:** Mapping to the STS Codes
* **sts\_name:** Mapping to the STS Names
* **icd10\_code:** Mapping to the ICD10 Codes
* **mild:** Flag for Mild CHD, used in automated complexity algorithm
* **moderate:** Flag for Moderate CHD, used in automated complexity algorithm
* **severe:** Flag for Severe CHD, used in automated complexity algorithm

## 1.2 Procedure Reference Table

The CHAANZ Procedure Reference Table (PRT), which is also centred on EPCC codes, is submitted separately, and comprises the following variables:

* **PROCEDURE\_IPCCC\_R:** The EPCC name of the procedure.
* **IPCCC\_CODE\_R:** The EPCC code for the procedure. Again, note that ‘IPCCC’ refers to an older version of the registry where the IPCCC long list was used. The current DRT, CHAANZ Registry and the PRT all use the EPCC list.
* **PROCEDURE\_STS\_R:** The PRT contains an output mapping to the STS procedure codes, this variable contains the STS procedure name that matches the EPCC Code
* **STS\_CODE\_R:** The PRT contains an output mapping to the STS procedure codes, this variable contains the STS procedure code that matches the EPCC Code, not a few options are not coded and suggest a manual review required.
* **PROC\_ELIGIBILITY:** Binary flag to denote that a procedure if present instantly qualifies a patient for inclusion in the CHAANZ Registry.

## 1.3 Summary of Eligibility Levels

For data collected from all sites, there are five eligibility flags for patient inclusion in the CHAANZ Registry:

* **Baseline:** Baseline Eligibility determined by the CHAANZELIGIBILITY variable in the DRT.
* **Baseline Procedure:** Baseline eligibility determine by the PROC\_ELIGIBILITY variable in the PRT.
* **Marfan:** Patients whose baseline flag = 0 and have diagnosis Marfan’s Syndrome (14.02.17), applying the rule type “Gene Specific”.
* **William:** Patients whose baseline flag = 0 and have diagnosis William’s Syndrome (14.02.30), applying the rule type “Gene Specific”.
* **LD:** Patients whose baseline flag = 0 and have diagnosis Loeys-Dietz Syndrome (14.04.85), applying the rule “Gene Specific”.

Patient who do not have a clear CHD diagnosis have a baseline flag equal to 0, but they may still be included in the CHAANZ Registry if Baseline Procedure, Marfan, William and/or Loeys-Dietz flag = 1.

For data collected at child sites, patients whose baseline flag = 1 are reviewed to assess three additions flags:

* **PFO:** Patients with baseline flag = 1 due to a single diagnosis of Patent foramen ovale (i.e. codes where the rule type = “PFO”).
* **PDA:** Patients with baseline flag = 1 due to a single diagnosis of diagnosis Patent arterial duct (i.e. codes where the rule type = “PDA”).
* **Valve:** Patients with baseline flag = 1 due to one or more valve-related diagnosis only (i.e. where the rule type = “Valve”).

# Eligibility at All Hospitals

## Baseline Eligibility

The variable CHAANZELIGIBILITY indicates a clear CHD diagnosis that instantly qualifies a patient for inclusion in the CHAANZ Registry. This ‘baseline’ level of eligibility is calculated and flagged with a variable called ‘baseline’ that equals 1 if a patient has at least one diagnosis where CHAANZELIGIBILITY = 1.

## Baseline Eligibility - Procedure

The variable PROC\_ELIGIBILITY indicates a clear CHD procedure that instantly qualifies a patient for inclusion in the CHAANZ Registry. This baseline-procedure level of eligibility is calculated and flagged with a variable called ‘baseline\_proc’ that equals 1 if a patient has at least one procedure where PROC\_ELIGIBILITY = 1.

## Genetic Eligibility

After the baseline eligibility, eligibility for three hereditary disorders that are sometimes associated with heart disease is calculated. These three hereditary disorders are Marfan’s syndrome (14.02.17), Williams syndrome (14.02.30), and Loeys-Dietz syndrome (14.04.85). For each condition, we created a separate binary flag variable (‘Marfan’, ‘William’, and ‘LD’) and searched the patient’s other diagnoses for the associated codes shown in Table 1 below. Each hereditary disease therefore has a distinct set of associated diagnostic codes. Presence of at least one of these associated codes with the hereditary syndrome qualifies a patient for inclusion in the CHAANZ registry and is indicated by the relevant flag being set to 1. The three hereditary diseases, their EPCC codes, and the associated codes that qualify patients for inclusion in the CHAANZ Registry are shown in Appendix 1.

## Final Eligibility

All five flags are considered when assessing the final eligibility of a patient. A patient is included in the Registry (and the data sent to Keysoft) only if at least one of the eligibility flags equals 1. For example, if a patient does not have a clear CHD diagnosis (i.e., CHAANZELIGIBILITY = 0 for all baseline diagnoses), they will still be included in the CHAANZ Registry if either their Baseline-procedure flag, Marfan flag, William flag and/or Loeys-Dietz flag equals 1. Each patient is therefore associated with five binary flag variables, together which determine final CHAANZ eligibility. All patients submitted to Keysoft must have at least one flag baseline, baseline\_proc, Marfan, Williams or LD equal to 1. The associated codes for each syndrome are detailed in Appendix 1.

## Flagging for Genetic Patients

To facilitate interrogation of data in the CHAANZ Registry, we create two additional binary flag variables. We called these flags ‘GENE\_PATIENT’ and ‘GENE\_ISOLATED’.

Each code that is a genetic condition has “GENE\_FLAG” = 1. Use this diagnosis level flag to create two patient level flags:

* GENE\_PATIENT – if equal to 1, then this patient has a genetic diagnosis in their list
* GENE\_ISOLATED – if equal to 1, then this patient has a genetic diagnosis as their only CHAANZ eligible diagnosis.

# 2.1 Additional rules for Paediatric sites

Data from each paediatric hospital will undergo another round of eligibility selection whereby some patients have their eligibility in the CHAANZ Registry revoked due to the presence of transient circulatory features, i.e. anatomical structures that are part of normal foetal development and may still appear in neonates but do not persist past 12 months.

To apply these additional rules for child sites, we create a follow-up file that contains all available encounter data. This includes any data that notes an interaction with the cardiology service and the date of the interaction, such as diagnosis or procedure records, clinic visits, cardiac, catheter or echocardiography visits, etc. We then calculate the age at each visit and delete all rows for age less than one year.

This file is used to create a list of patient IDs, where evidence is available to indicate a continuation of patient follow-up with a cardiac service at greater than one year of age.

## PFO, PDA, and VALVE rules

The DIAGNOSIS Table is used to create two files. The first file contains a list of unique patient IDs for all patients with at least one diagnosis associated with a RULE\_TYPE of either PFO, PDA or VALVE.

The second file comprises a list of patients that have at least one diagnosis where CHAANZELIGIBILITY = 1. Diagnoses where RULE\_TYPE = GENE\_SPECIFIC are excluded from this file because these are dealt with separately.

We then compare these two files to create a new list of patients that have at least one diagnosis where RULE\_TYPE is either PFO, PDA or VALVE, but there is no other diagnosis for which CHAANZELIGIBILITY = 1. These patients are individuals that are initially considered for inclusion in the Registry but because their qualifying diagnosis is a transient circulatory feature present in many infants, their participation in the Registry can be revoked.

To determine whether Patients with PFO, PDA, or VALVE diagnoses (and no other CHD diagnosis) are eligible for the Registry we consult the follow-up file mentioned above. The follow-up file contains all data after one year of age for all sources. If an ID is found in this follow-up list, we assume that the patient has had follow-up with the cardiac service after one year of age. Patients who have had follow-up remain in the Registry while patients that have no evidence of follow-up after 1 year are removed from Registry participation.

**Appendix 1. Associated codes for hereditary diseases of interest to the CHAANZ Registry.**

**Marfan’s associated codes:**

* ascending aorta dilatation
  + Aortic abnormality, Ascending aorta dilation, - 09.16.09
  + Abnormality of aorta: acquired, Ascending aorta dilation: acquired - 10.14.40
* aortic aneurysm
  + Abnormality of aorta: acquired, Aortic aneurysm, Ascending aorta aneurysm, - 10.14.42
  + Abnormality of aorta: acquired, Aortic aneurysm, Descending aorta aneurysm, - 10.14.43
  + Abnormality of aorta: acquired, Aortic aneurysm, Abdominal aorta aneurysm, - 10.14.44
  + Abnormality of aorta: acquired, Aortic aneurysm, - 10.14.50
  + Abnormality of aorta: acquired, Aortic dissection - 10.14.51
  + Descending aorta dilation, - 09.28.16
* mitral valve prolapse
  + Mitral valvar abnormality, Mitral valvar prolapse, - 06.02.35

**William’s Syndrome a*ssociated codes:***

* supra aortic stenosis
  + Aortic abnormality, Supravalvar aortic stenosis, 09.16.00
  + Abnormality of aorta: acquired, Supravalvar aortic stenosis: acquired, 10.14.77
* pulmonary artery stenosis
  + Pulmonary arterial abnormality, Pulmonary arterial stenosis, 09.10.01
  + Pulmonary arterial abnormality, Pulmonary arterial stenosis, Peripheral pulmonary arterial stenoses or hypoplasia: at-beyond hilar bifurcation, 09.10.06
  + Pulmonary arterial abnormality, Pulmonary arterial stenosis, Central pulmonary arterial stenosis or hypoplasia: proximal to hilar bifurcation, 09.10.07
  + Pulmonary arterial abnormality, Pulmonary arterial stenosis, Right pulmonary arterial stenosis, 09.10.25
  + Pulmonary arterial abnormality, Pulmonary arterial stenosis, Left pulmonary arterial stenosis, 09.10.26
* middle aortic syndrome
  + Descending-abdominal aorta hypoplasia (middle aortic syndrome), - 09.29.16

**LD Syndrome a*ssociated codes:***

* + ascending aorta dilatation Aortic abnormality, Ascending aorta dilation, - 09.16.09
  + Abnormality of aorta: acquired, Ascending aorta dilation: acquired - 10.14.40
* aortic aneurysm
  + Abnormality of aorta: acquired, Aortic aneurysm, Ascending aorta aneurysm, - 10.14.42
  + Abnormality of aorta: acquired, Aortic aneurysm, Descending aorta aneurysm, - 10.14.43
  + Abnormality of aorta: acquired, Aortic aneurysm, Abdominal aorta aneurysm, - 10.14.44
  + Abnormality of aorta: acquired, Aortic aneurysm, - 10.14.50
  + Abnormality of aorta: acquired, Aortic dissection - 10.14.51
  + Descending aorta dilation, - 09.28.16
* mitral valve prolapse
  + Mitral valvar abnormality, Mitral valvar prolapse, - 06.02.35 (CHD = 1)