

**The data entry manual is designed to provide a clear definition for each variable collected and the options for each variable**

## **SAMPLE Data Entry Manual for the vEDS Project**

### **Subject ID**

Each study participant (or patient) should have a single 'Subject ID.' Please use an abbreviation of your institution name with a chronological number attached to it. For example, at UCLA we would use the following Subject IDs: UCLA001, UCLA002, UCLA003, etc.

### **Patient Demographics**

#### **1. Sex**

Select Male or Female.

#### **2. Race**

Select from the following: "Caucasian," "African American," "Hispanic White," "Hispanic not white" or "Other." You may enter free text in the "Other" option. If race is unknown, please enter "unknown" into the "Other" box.

#### **3. Height**

Enter the most recent height of the patient (in centimeters). Please do not enter the units in the form. An example of the correct entry is "170."

#### **4. Weight**

Enter the most recent weight of the patient (in kilograms). Please do not enter the units in the form. An example of the correct entry is "80."

#### **5. Age at Diagnosis**

Enter the age of the patient (in years) at the time of diagnosis. Please do not enter the units in the form. An example of the correct entry is "12."

#### **6. Method of Diagnosis**

Select from "Clinical Criteria", "Genetic Testing", or "Skin Biopsy" as the highest level of diagnosis that the patient has received. For example, if the patient met clinical criteria, but then had genetic testing done to confirm the diagnosis, enter "genetic testing".

#### **7. COL3A1 gene mutation**

Enter the specific COL3A1 gene mutation found in the patient. An example of the correct entry for this form is "c.746G>A"

#### **8. Protein mutation**

Enter the specific protein mutation found in the patient. An example of the correct entry is "p.(Gly249Asp)"

## 9. Family History

Select from “History of vEDS”, “Sudden Death”, “Early CV Disease”, “none of the above” or “Unknown”. For Mother, Father, Sibling, and Extended. If multiple siblings or multiple extended family members exist, enter in the “Describe” box with relation such as “Sister: history of vEDS, Brother: Sudden death”

## 10. Life Status

Select from “alive” or “dead” as of most recent appointment or contact.

### 10a. Age Today

Enter patient’s current age in years. Please do not enter the units in the form. An example of the correct entry is “70.”

### 10b. Age at death

Enter patient’s age at the time of death in years. Please do not enter the units in the form. An example of the correct entry is “70.”

### 10c. Cause of death

Enter the cause of death. If unknown, enter “unknown”

## 11. Autopsy performed

Select yes or no if autopsy was performed.

### 11a. Autopsy findings

Enter autopsy findings as free text. Be as specific as possible, attach de-identified report if possible.

## Clinical Criteria

**12. Easy bruising; Arterial, Intestinal, or Uterine fragility; Thin, translucent skin; Characteristic facial features; Small joint hypermobility; Skin hyperextensibility; Spontaneous pneumothorax/hemothorax; Tendon or muscle rupture; Early-onset varicose veins; Talipes equinovarus (clubfoot); Carotid-cavernous fistula**

Select “Yes”, “No” or “Unknown” for each of the above listed variables. If patient exhibits other relevant clinical criteria or has specific note about criteria listed above, enter in “describe other”.

## Comorbidities

**13. Smoking History; Hypertension; Hyperlipidemia; Diabetes Mellitus; Previous MI; Coronary Artery Disease; Stroke; TIA; Polycystic Ovarian Syndrome; Deep Vein Thrombosis; Pulmonary Embolism**

Select “Yes”, “No” or “Unknown” for each of the above listed variables. If patient has other comorbidities not listed, or has specific note about a comorbidity listed above, enter in “describe other”.

## Pregnancy and GI Complications

**14. Gravida, Parity, Spontaneous Abortions, Termination of Pregnancy, Living children**

Enter number of each event above. If patient is female but did not experience an event, please enter “0”. If patient is male, please leave fields blank.

## 15. Diagnosis in Relation to Pregnancy

Enter whether patient was diagnosed before, during, or after first pregnancy. If patient was diagnosed during pregnancy, please then enter time at which diagnosis was made, in weeks of gestation.

**16. Pregnancy complications**

Select “yes” or “no” if there were complications with the pregnancy or birth. Describe the complications in the “describe” box below.

**17. GI involvement**

Select “yes” or “no” if the patient has experienced GI involvement in their lifetime (bowel rupture, etc.).

**17a. Type of Initial GI presentation**

Select whether or not the initial GI presentation was “elective”, “emergent”, or “unknown”.

**17b. Known Diagnosis at Initial GI presentation**

Select “yes” or “no” or “unknown” if a diagnosis was known at the initial presentation for GI involvement.

**17c. Age at Initial GI presentation**

Enter patient’s age at the time of GI involvement. Please do not enter the units in the form. An example of the correct entry is “20.”

**Pneumothorax and Hemothorax****18. Pneumothorax (PTX)**

Select “yes” or “no” if the patient has experienced a pneumothorax in their lifetime.

**18a. Age at time of PTX**

Enter patient’s age at the time of PTX in years. Please do not enter the units in the form. An example of the correct entry is “20.”

**18b. Known Diagnosis at time of first PTX**

Select “yes” or “no” or “unknown” if a diagnosis was known at the initial presentation for PTX.

**18c. Hospital length of stay**

Enter the length of hospital stay for PTX in days. Please do not enter the units in the form. An example of the correct entry is “20.”

**18d. Management**

Describe how the PTX was managed. Be as detailed as possible.

**18e. Recurrent PTX**

Enter the number of recurrent PTX that the patient endured.

**18f. Management**

Describe how the recurrent PTX was/were managed. Be as detailed as possible.

## 19. Hemothorax (HTX)

Select “yes” or “no” if the patient has experienced a hemothorax in their lifetime.

### 19a. Age at time of HTX

Enter patient’s age at the time of HTX in years. Please do not enter the units in the form. An example of the correct entry is “20.”

### 19b. Known Diagnosis at time of first HTX

Select “yes” or “no” or “unknown” if a diagnosis was known at the initial presentation for HTX.

### 19c. Hospital length of stay

Enter the length of hospital stay for HTX in days. Please do not enter the units in the form. An example of the correct entry is “20.”

### 19d. Management

Describe how the HTX was managed. Be as detailed as possible.

### 19e. Recurrent PTX

Enter the number of recurrent HTX that the patient endured.

### 19f. Management

Describe how the recurrent HTX was/were managed. Be as detailed as possible.

## 20. Thoracic Aortic Surgery

Select “yes” or “no” if the patient has undergone thoracic aortic surgery at any time in their life. If yes, please fill out surgical details in questions 23 to 45.

## 21. Abdominal Aortic Surgery

Select “yes” or “no” if the patient has undergone abdominal aortic surgery at any time in their life. If yes, please fill out surgical details in questions 23 to 45.

## Initial Vascular Presentation

**(Please add all events under the same patient ID number.)**

## 22. Type of Initial Vascular Presentation

Select whether or not the initial GI presentation was “elective”, “emergent”, or “unknown”.

### 22a. Known Diagnosis at time of first vascular presentation

Select “yes” or “no” or “unknown” if a diagnosis was known at the initial vascular presentation.

### 22b. Age at time of initial vascular presentation

Enter patient’s age at the time of initial vascular presentation in years. Please do not enter the units in the form. An example of the correct entry is “20.”

### 22c-f. Arterial Involvement

Select the artery involved in the initial vascular presentation. For type, select “dissection”, “aneurysm”, “dissection and aneurysm”, “rupture”, or “other involvement not listed”. If “other” is selected, please enter the type of involvement in the box below.

## **Surgery 1**

**(Please add all surgeries under the same patient ID number)**

### **23. Vascular procedure type**

Select “open”, “endo”, “endo converted to open”, or if the vascular involvement was “medically managed.”

### **24. Endo Procedure Type**

Select “Diagnostic”, “embolization”, “angioplasty”, or “stent”.

### **25. Indication for Procedure**

Enter why the procedure was indicated in this patient.

### **26. Access**

Select whether the procedure was performed “percutaneous” or as a “cut down”

### **27. Largest Sheath Inserted**

Enter the largest sheath size entered, with units. An example of a correct entry for this field is “6F”.

### **28. Closure Method**

Enter how the entry site was closed for this procedure.

### **29. Cell Saver used**

Select “yes” or “no” or “unknown” as to whether or not Cell Saver was used during this procedure.

#### **29a. Cell Saver Quantity used**

If 29 was entered “yes”, enter the amount of Cell Saver used in this procedure, in mL. Please do not enter the units in the form. An example of the correct entry is “20.”

### **30. Blood Transfusion**

Select “yes” or “no” or “unknown” as to whether or not the patient received a blood transfusion during this procedure.

#### **30a. PRBC Quantity, FFP Quantity, Platelets Quantity, Cryo Quantity, Fibrinogen Quantity**

Enter the amount of the listed products in mL. Please do not enter the units in the form. An example of the correct entry is “20.”

### **31. Percutaneous Access Site Complications**

Enter any complications experienced with the access site. If no complications occurred, please enter “none”.

### **32. Wound Complications**

Select “infection”, “hematoma”, “lymph leak”, or “none” to indicate if a wound complication occurred.

#### **32a. Management of Wound Complications**

Enter how the wound complications were subsequently managed.

**33. Abdominal Wound Dehiscence**

Select “yes” or “no” or “unknown” as to whether or not there was dehiscence of an abdominal wound. If “yes”, please describe further how it was managed in the field below.

**34. In Hospital Mortality**

Select “yes” or “no” as to whether or not the patient passed away while in the hospital.

**35. Mechanical Ventilation >24 hours**

Select “yes” or “no” as to whether or not the patient required to be on a mechanical ventilator for longer than 24 hours.

**35a. Days on Mechanical Ventilation**

Enter the number of days the patient required mechanical ventilation. Please do not enter the units in the form. An example of the correct entry is “20.”

**36. Pneumonia**

Select “yes” or “no” as to whether or not the patient experienced a pneumonia while in the hospital.

**37. Stroke**

Select “yes” or “no” as to whether or not the patient experienced a stroke while in the hospital. Please fill out the field below with further details.

**38. Acute Renal Failure (ARF)**

Select “yes” or “no” as to whether or not the patient experienced acute renal failure while in the hospital.

**38a. Need HD**

Select “yes” or “no” as to whether or not the patient required hemodialysis as a result of acute renal failure while in the hospital.

**38b. Resolved**

Select “yes” or “no” or “unknown” as to whether or not the ARF was resolved.

**39. Deep Vein Thrombosis (DVT)**

Select “yes” or “no” as to whether or not the patient experienced deep vein thrombosis while in the hospital.

**40. Myocardial Infarction (MI)**

Select “yes” or “no” as to whether or not the patient experienced a myocardial infarction while in the hospital.

**41. Troponin Leak**

Select “yes” or “no” as to whether or not the patient experienced a troponin leak while in the hospital.

**42. Atrial Fibrillation (A-Fib)**

Select “yes” or “no” as to whether or not the patient experienced atrial fibrillation while in the hospital.

**43. OR Takeback**

Select “yes” or “no” as to whether or not the patient was taken back into the OR for further surgery while in the hospital.

**44. Total days in hospital**

Enter the total number of days spent in the hospital. Please do not enter the units in the form. An example of the correct entry is “20.”

**44a. Days in ICU**

Enter the total number of days the patient spent in the ICU. Please do not enter the units in the form. An example of the correct entry is “20.”

**44b. Discharged to**

Select “home”, “SNF” (specialized nursing facility), “rehab”, or “unknown” as the place the patient was discharged to.

**45. Readmission**

Select “yes” or “no” as to whether or not the patient was readmitted within 30 days of discharge. Please describe further in the field below.

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