The purpose of the National Familial Hypercholesterolaemia Registry is to collate data to facilitate clinical service planning and to inform clinical best practice. The Registry will also enable research on aggregated data and the identification of eligible volunteers for clinical trials.

The National fH Registry

USER MANUAL

fH Australasia Network

everyone deserves a better chance

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Need Help

Contact National Coordinator: Dr Jing Pang
Phone: (08) 9224 0318
Email fhwa@health.wa.gov.au

Resources

- The National Familial Hypercholesterolaemia Registry - an introduction (5:51 min video) (https://www.youtube.com/watch?v=kIkGys77fHw)
  N.B. The user interface has been updated since this video was created, but it is still useful to demonstrate general concepts.

- National FH Registry (https://fhregistry-international.com/)

- National FH Registry - Username/Password screen (https://fhregistry-international.com/rdrf/login?next=/rdrf/router/)

- National FH Registry – Charter, Protocol and Guidelines (http://www.athero.org.au/fh/wp-content/uploads/National-FH-Registry-–-Charter-Protocol-and-Guidelines.pdf)

- Registry Brochure (http://www.athero.org.au/fh/wp-content/uploads/FH-Registry-Brochure_v4.pdf)

- Sample Consent Form (http://www.athero.org.au/fh/wp-content/uploads/Sample-Consent-Form.pdf)

- Test site (https://rdrf.ccgapps.com.au/ophg/login?next=/ophg/router/)
  - Username: fhcurator
  - Password: FHcurator1*

Abbreviations used in the FH Registry

DLCNS – Dutch Lipid Clinic Network Score

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Eligibility Criteria for Inclusion in the FH Registry

- Adult INDEX with:
  - Dutch Lipid Clinic Network Score (DLCNS) ≥ 6 (ie. **probable** or **definite** FH)
  - Genetic mutation that is causative of FH
  - Familial elevation of Lp(a) *– this will be a subset of the larger FH registry*
    - Defined as Lp(a) > 0.5g/L with a DLCNS ≥ 3

- Child/adolescent INDEX with:
  - Classification of **highly probable** FH
    - LDL-cholesterol > 5.0 mmol/L
  - Classification of **probable** FH
    - LDL-cholesterol > 4.0 mmol/L with a family history of premature heart disease
    - LDL-cholesterol > 4.0 mmol/L with hypercholesterolaemia or tendon xanthoma or arcus in the first-degree relative
  - Genetic mutation that is causative of FH
  - Familial elevation of Lp(a) *– this will be a subset of the larger FH registry*
    - Defined as Lp(a) > 0.5g/L with a family history of premature heart disease

- **All** relatives of the above including those who have tested negative
  - Classified according to age- and gender-specific LDL-cholesterol cut-offs (link)

Also record Familial Combined Hyperlipidaemia (FCHL) if they coincide with the above.

Hypertension Criteria

Hypertension is defined as:

- Systolic blood pressure > 140 mmHg **or**
- Diastolic blood pressure > 90 mmHg

Hypertriglyceridaemia Criteria

Hypertriglyceridaemia is defined as:

- Triglyceride > 10 mmol/L **or**
- Mutations in LPL, APOA5, APOC2, LMF1, GPIHBP1, APOE, APOC3 **or**
- Genetic risk score indicative of hypertriglyceridaemia

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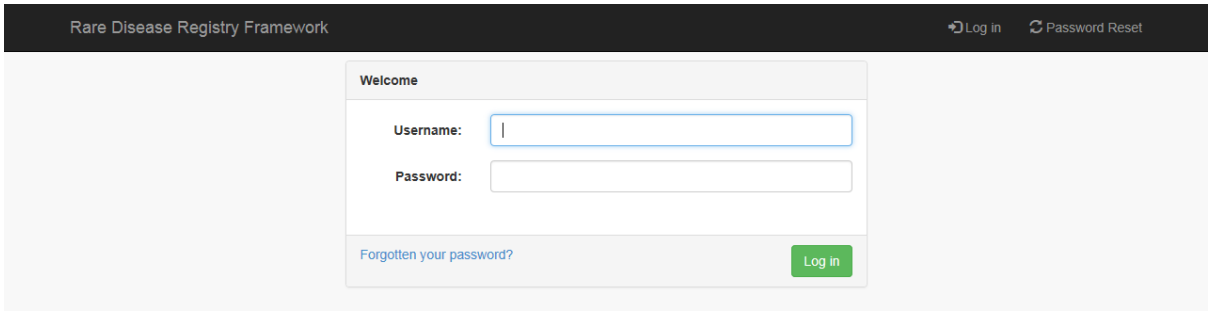
Logging In

Go to https://fhregistry-international.com/

Click **Registry**



Suggestion: Bookmark/add to favourites the page for later use.

- Enter your username and password
- Click **Log in**. The Patient List screen will appear.

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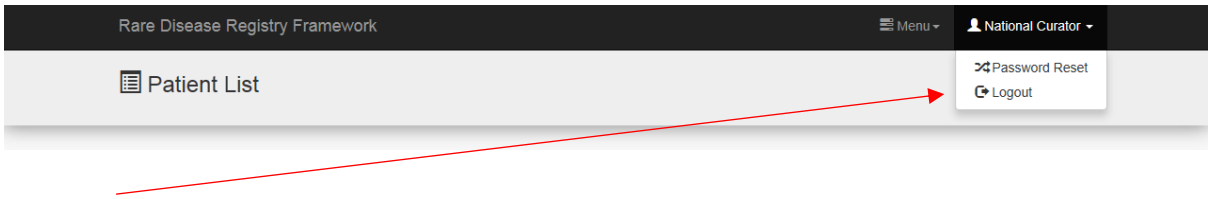
Logging Out



Click **Logout**

Password Requirements

The FH Registry uses the following password policy:

- Must be at least 8 characters long
- Must contain at least one of each of the following
    - Uppercase letter
    - Lowercase letter
    - Number
    - Special character (!, @, #, $, %, ^, &, *).

Password Deactivation

Your password will be deactivated after three months of non-use.

You will need to email the National Coodinator to arrange reactivation.

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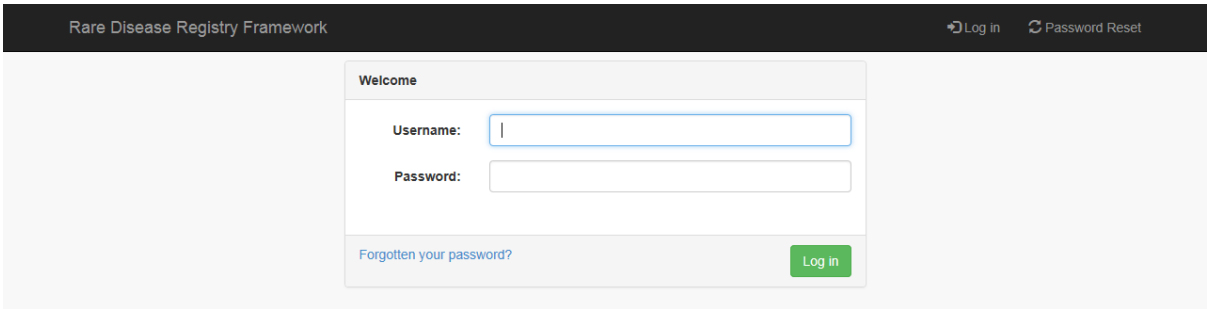
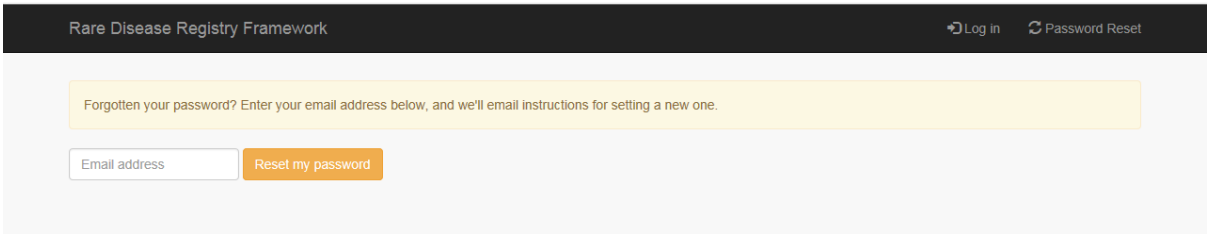
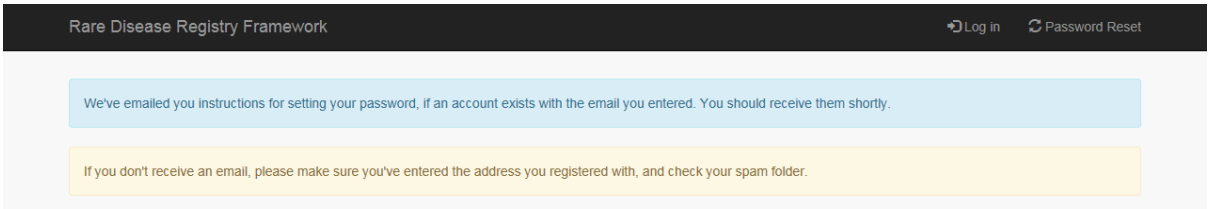
Forgotten your Password?



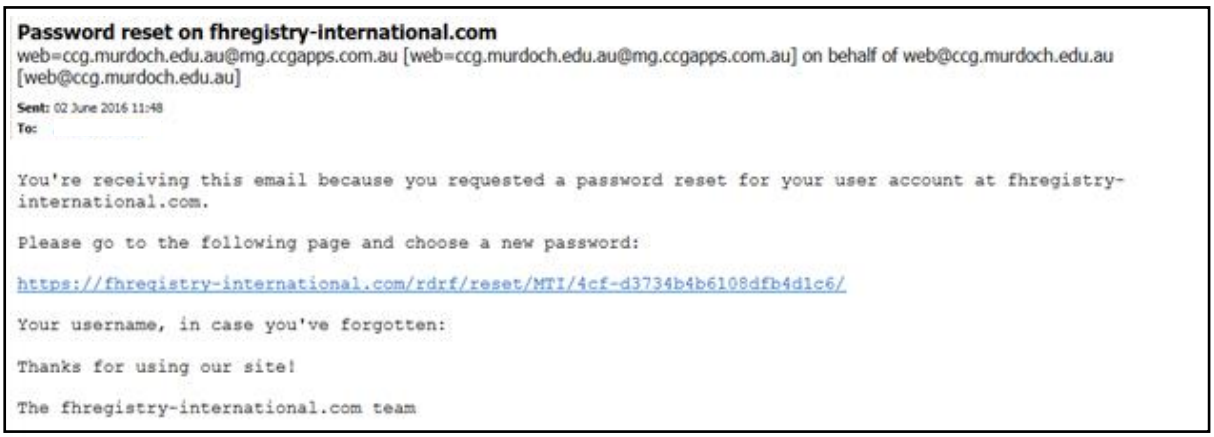
- If you have forgotten you password, click **Forgotten your password?**



- Enter your email address which was registered with the Registry
- click **Reset my password**



- Go to your email account



- You will receive this email. Click the blue link.

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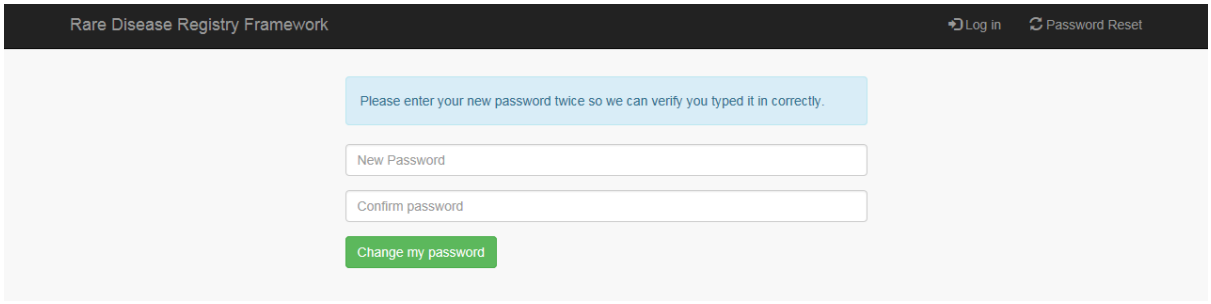
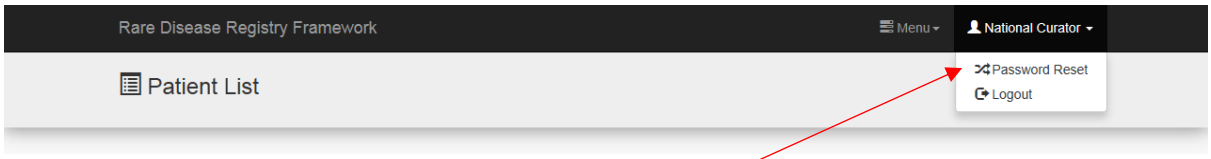


- Enter your new password and confirm
- Click **Change my password**

- Click **Log in** to take you to the Log in screen



N.B. Password can be reset at any time from the dropdown menu.

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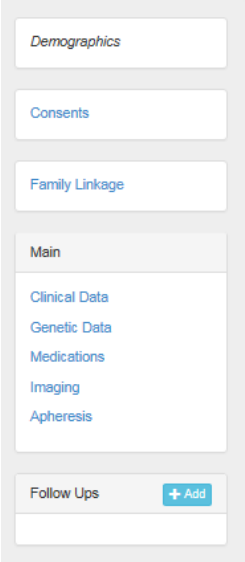
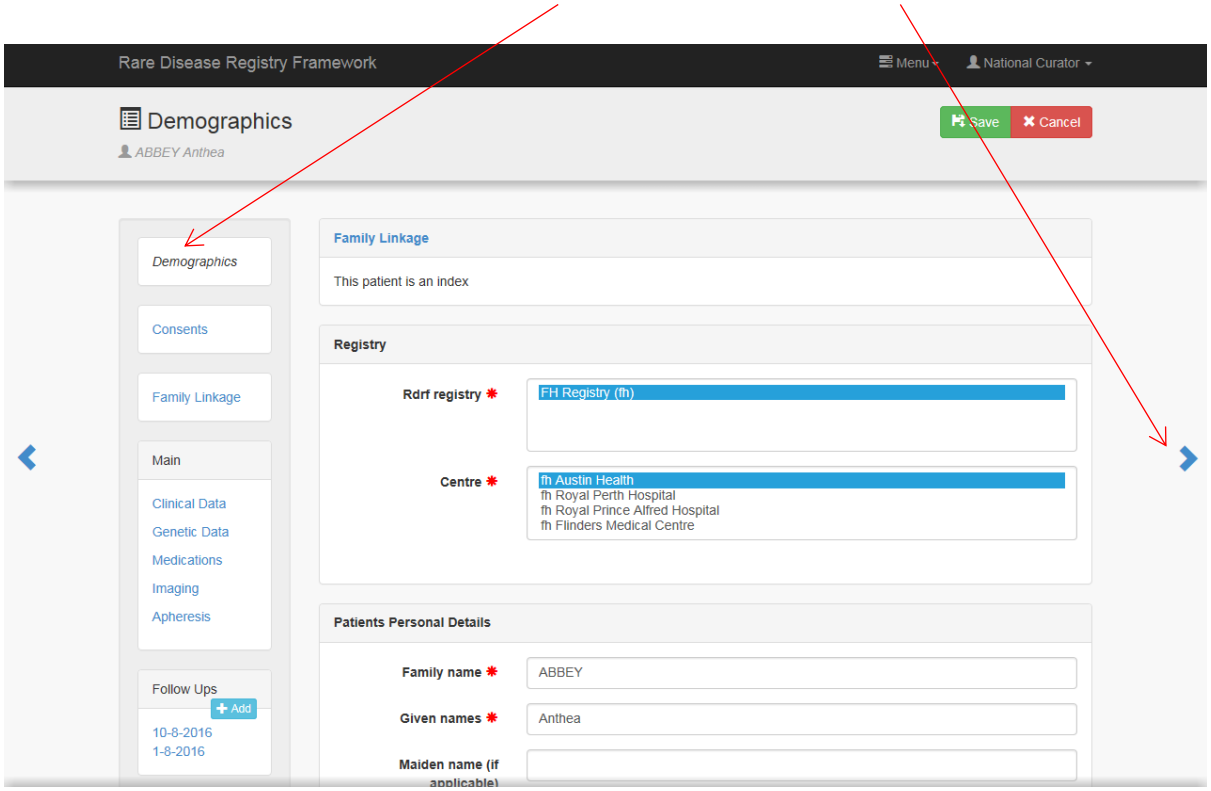
Structure of the FH Registry



All patient records have the following modules:

- Demographics (Screen shot – Demographics)
- Consents (Screen shot – Consents)
- Family Linkage (Screen shot – Family Linkage)
- Clinical Data (Screen shot – Clinical Data)
- Genetic Data (Screen shot – Genetic Data)
- Medications (Screen shot – Medications)
- Imaging (Screen shot – Imaging)
- Apheresis (Screen shot – Apheresis)
- Follow Up (multiple forms) (Screen shot – Follow Up)

You can move between the modules via the menu on the left or the next/previous arrows.



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Relationship between Index and Relative

The **INDEX** is created on the **Patient List** page by the **+ Add Patient** button

RELATIVES are created on the **Index's demographic page** via the **Patient Relative** field

Follow up
Apheresis
Imaging **
Genetics Data
Medications
Clinical Data
Consents
Demographics
INDEX
Relative 1
Relative 2

Follow up
Apheresis
Imaging *
Genetics Data
Medications
Clinical Data
Consents
Demographics
Relative 1

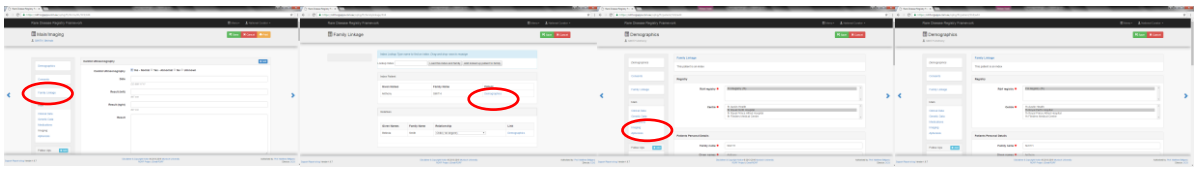
Follow up
Apheresis
Imaging
Genetics Data
Medications
Clinical Data
Consents
Demographics
Relative 2

Move between modules using the left hand menu or **Next/Previous** arrows

Move between **Relatives** and the **Index** via the Family Linkage

If you are on Relative 1's Imaging page* and want to look at the Index's imaging**:

- Click Family Linkage link (on Relative 1 imaging page)
- Click Index's Demographics link (on the Family Linkage page)
- Click Imaging link (on Index's Demographics page)
- Index's Imaging page



Relative 1 - Imaging | Family Linkage | Index Demographics | Index - Imaging

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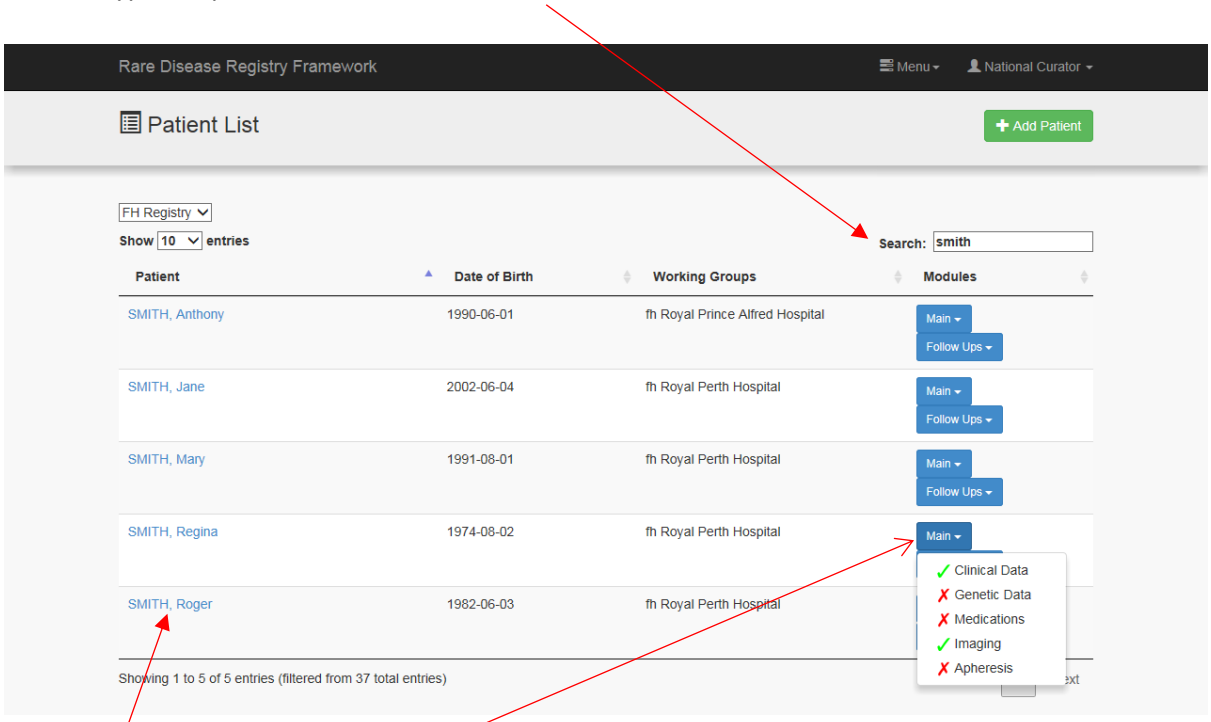
Patient List (screen shot – Patient List)

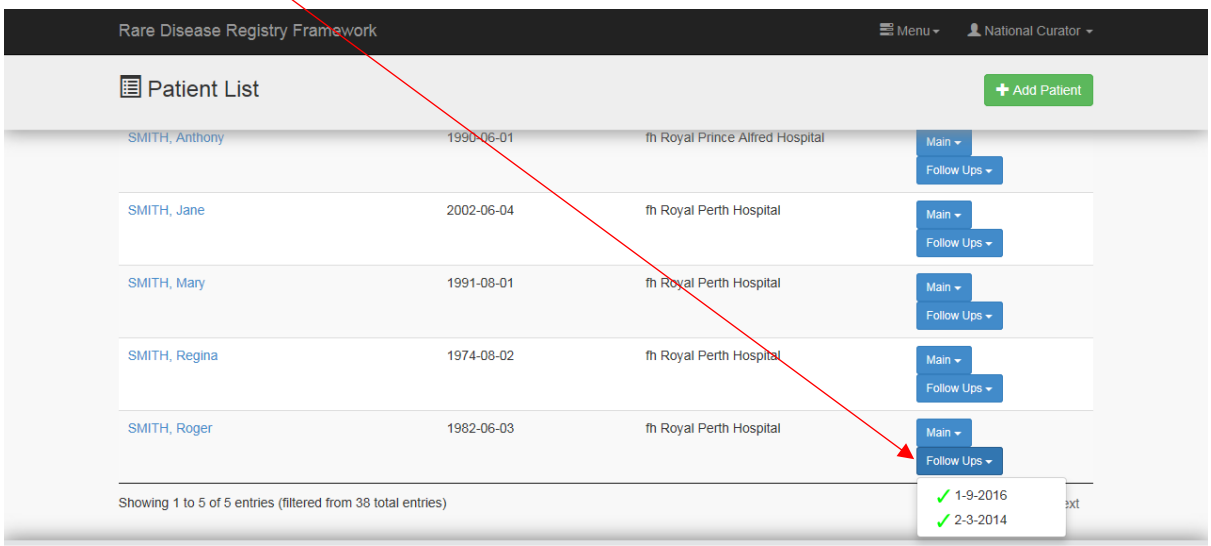
If the patient has already been entered in the FH Registry:

- Type the patient's name into the **Search** function



One or a list of names will then appear:

- Click the required name to take you to the patient's **Demographics** page
- Click **Main** and a dropdown menu will appear and you can click a specific module
  N.B. The ✔ tick indicates the module has had data entered and the ✘ cross indicates the module has not had data entered.
- Click **Follow Ups** and a dropdown menu will appear and you can click a specific follow up module



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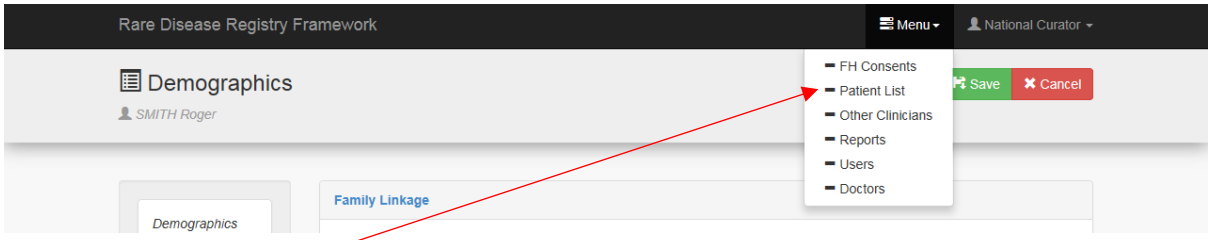

N.B. The **Patient List** can be accessed from any page using the dropdown menu.

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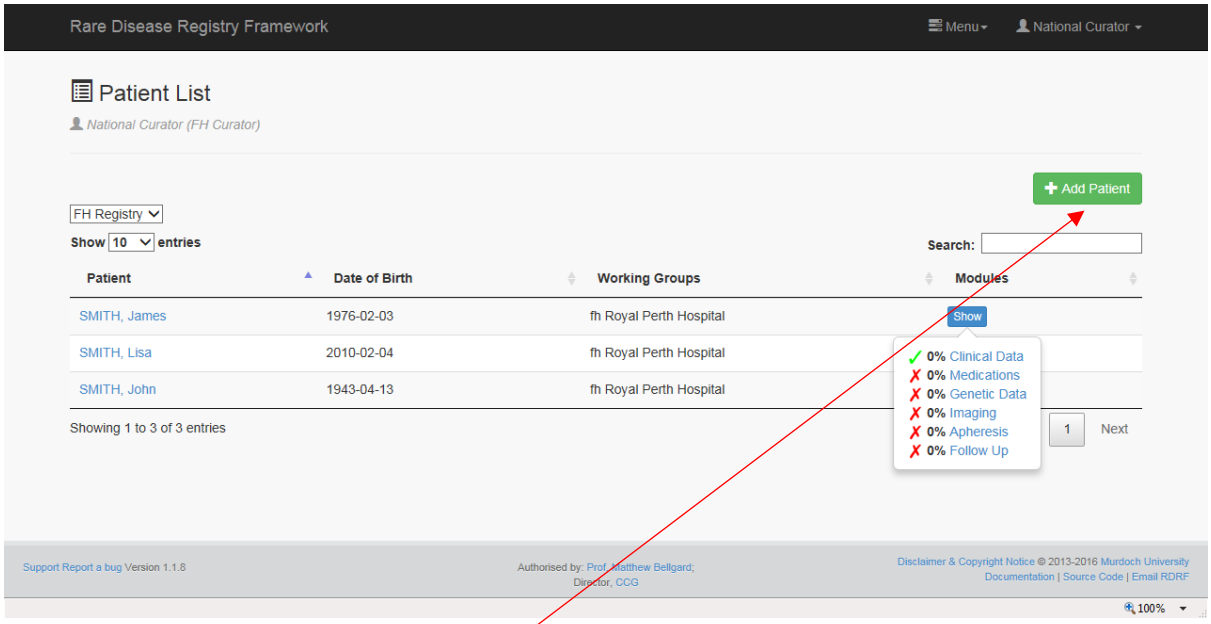
Add an Index Patient

The first person diagnosed with FH in the family is called the Index patient. All Relatives are linked to this Index.



An Index can only be added on the Patient List page.

To add a new Index patient:

- Click **+ Add Patient**
- A blank Demographics page will appear

N.B. DO NOT use the **+ Add Patient** button if you are adding a relative.

Relatives are entered on the Index's Demographics page. See:

- Add a **Relative**
- Change an incorrectly entered **Index** patient to a **Relative**

RETURN TO MENU

Demographics page (screen shot – Demographic page)

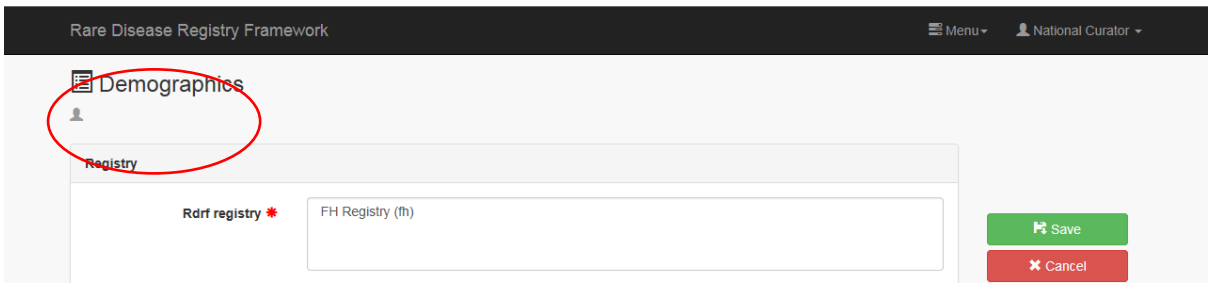
Related topics:

- Registry and Centre
- Date Function
- Patient Address
- Add Patient Doctor
    - Add Doctor's Details to the Database
    - Add Patient Doctor
- Add a Relative
    - Enter a Relative in a Different Centre to the Index
    - View Family Members in Different Centre
    - Delete a Relative
    - Delete an Index
- Pedigree

- Change an incorrectly entered Index patient to a Relative
- Change the Index

When the **+ Add Patient** button (to add a new Index) on the **Patient List** page is clicked, a blank **Demographics** page will appear.

- Complete all the required fields as indicated by *
- Click **Save**



Demographics page BEFORE saving

- No name beside person icon

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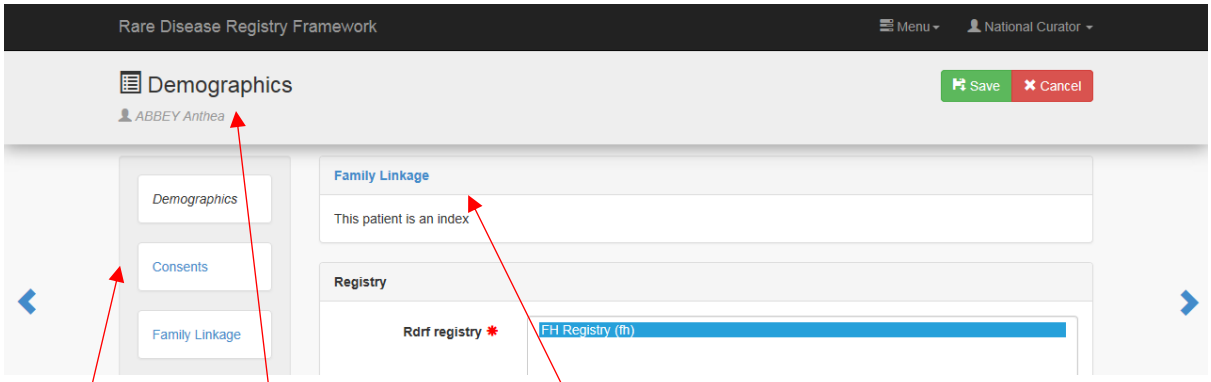


The **Index**'s Demographic page AFTER saving:

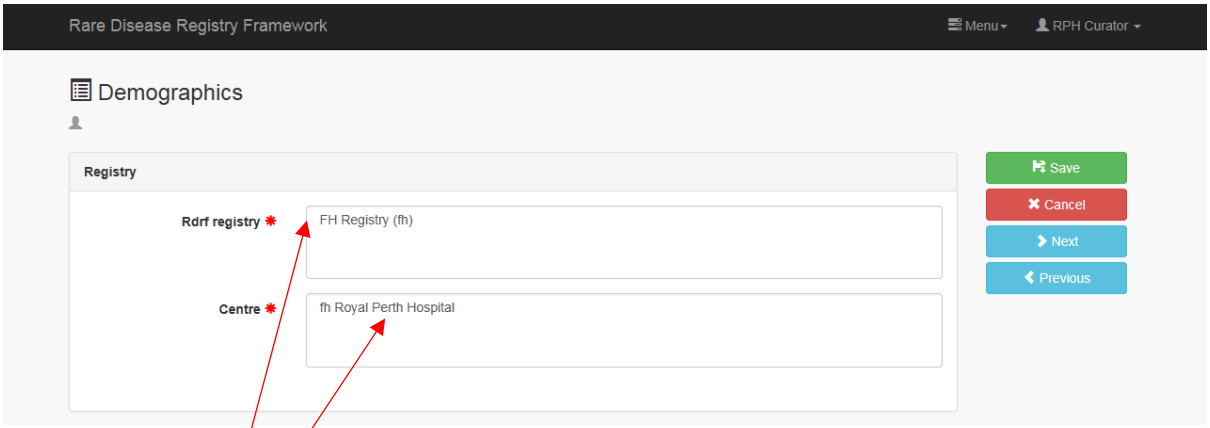
- A set of modules has been created for this Index
- Patient name now next to person icon
- Family Linkage and **This patient is an index** now visible

The **Relative**'s Demographic page AFTER saving:

- A set of modules has been created for this relative
- Patient's name appears next to person icon
- **Family Linkage** and **This patient is a relative of index…..**
- Clicking the Index's name will take you to the Index's Demographics page

RETURN TO MENU

Registry and Centre



- Click **FH Registry (fh)**
  N.B. Provision has been made for multiple registries to be linked in the future.
- Click **Centre** the patient is attending
  N.B. One or more Centres may be visible depending on your permissions.

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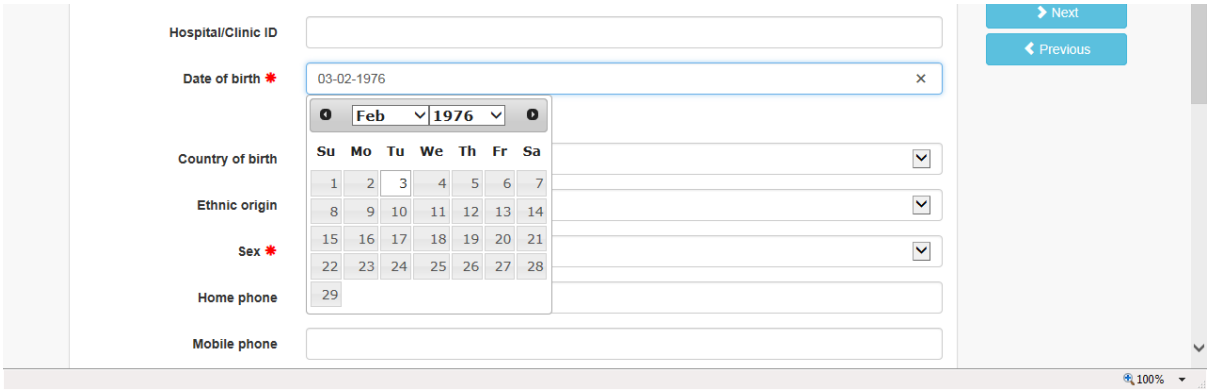
Date Function

The date can be entered manually as 3-2-1976 or 03-02-1976.

The dropdown function is based on a calendar:

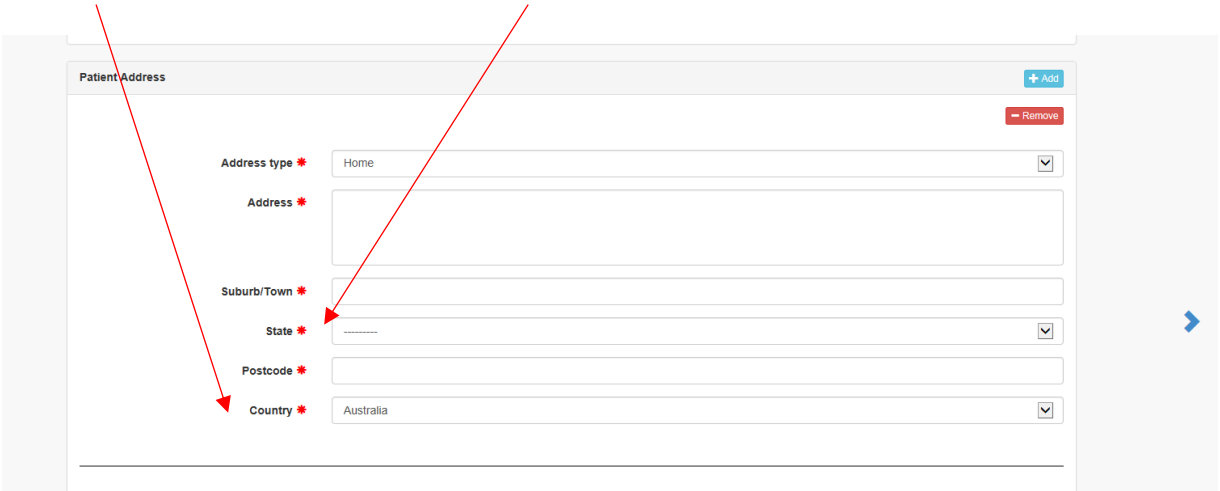
- The year must be selected first (the months for the selected year will then be loaded)
- Then the month selected (the days for the selected month will then be loaded)
- Finally the day selected

Once the day is selected the date will be entered automatically.



Patient Address

The **Country** must be selected before the **State** can be selected.



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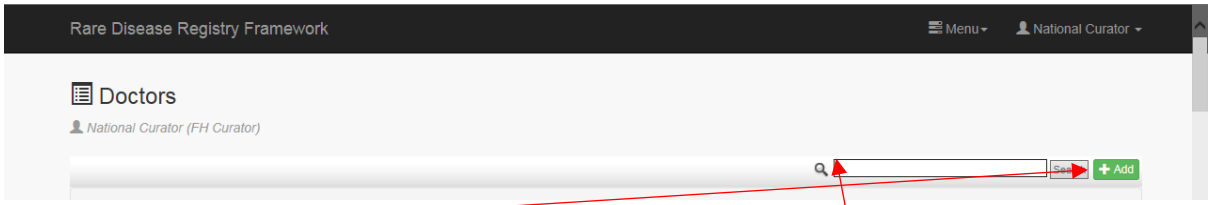
Add Patient Doctor

Add **Patient Doctor** occurs in two steps:

1. Doctor's details are added to the database. Details are only added once.
2. Add **Patient Doctor** on the patient's demographics page

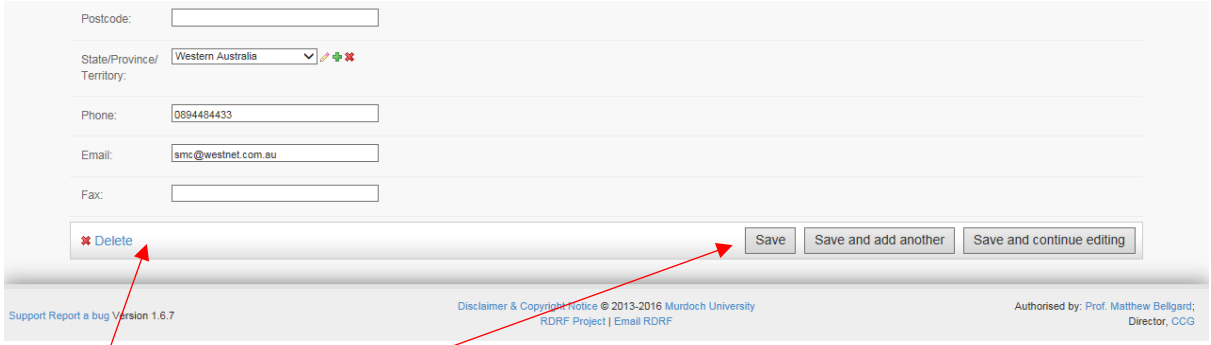
Add Doctor's Details to the Database

- Click Menu
- Click **Doctors** in the dropdown menu



- Click **+ Add** button

N.B. You are able to search for a doctor and change details via the search function.



- Enter doctor's details. Required fields are bolded.
- Click **Save**

N.B. The doctor's details can be deleted by clicking **Delete.**

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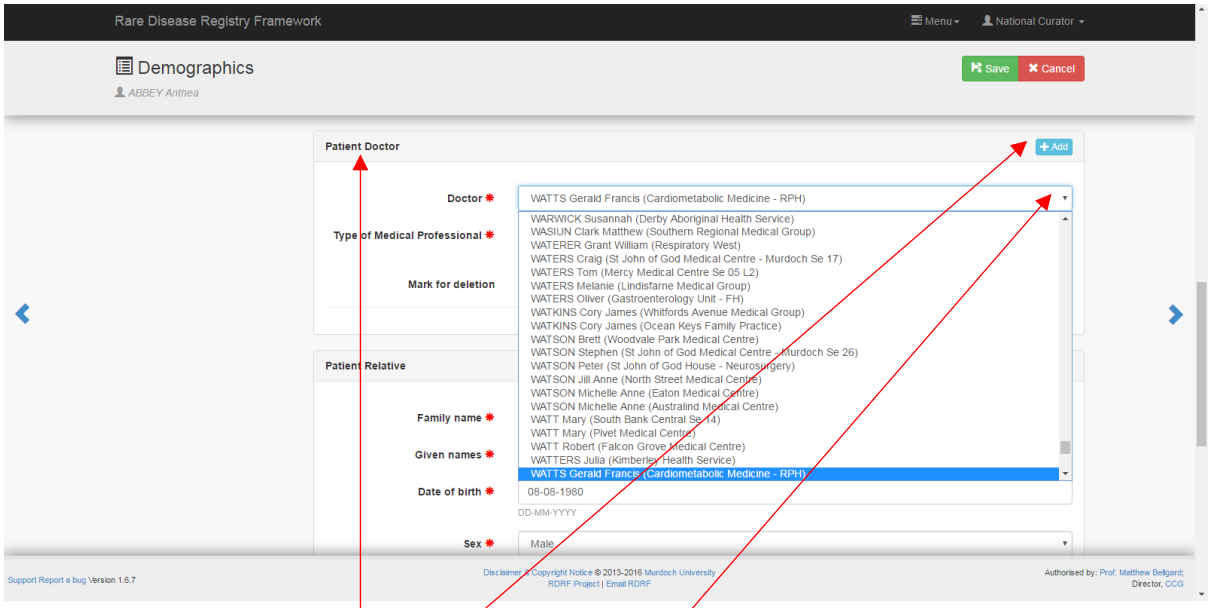
Add Patient Doctor

Once added to the database the doctor's name will automatically appear in the **Doctor** dropdown menu.



- Go to the patient's **Demographics** page
- Scroll down to **Patient Doctor**
- Click **+ Add** button
- Click the dropdown menu next to **Doctor.** Click the doctor's name or start typing the doctor's name, the doctor's name will appear in the box.
- Click the dropdown menu next to **Type of Medical Professional.** Click **Type of Medical Professional** and the details will appear in the box.
- Click **Save**

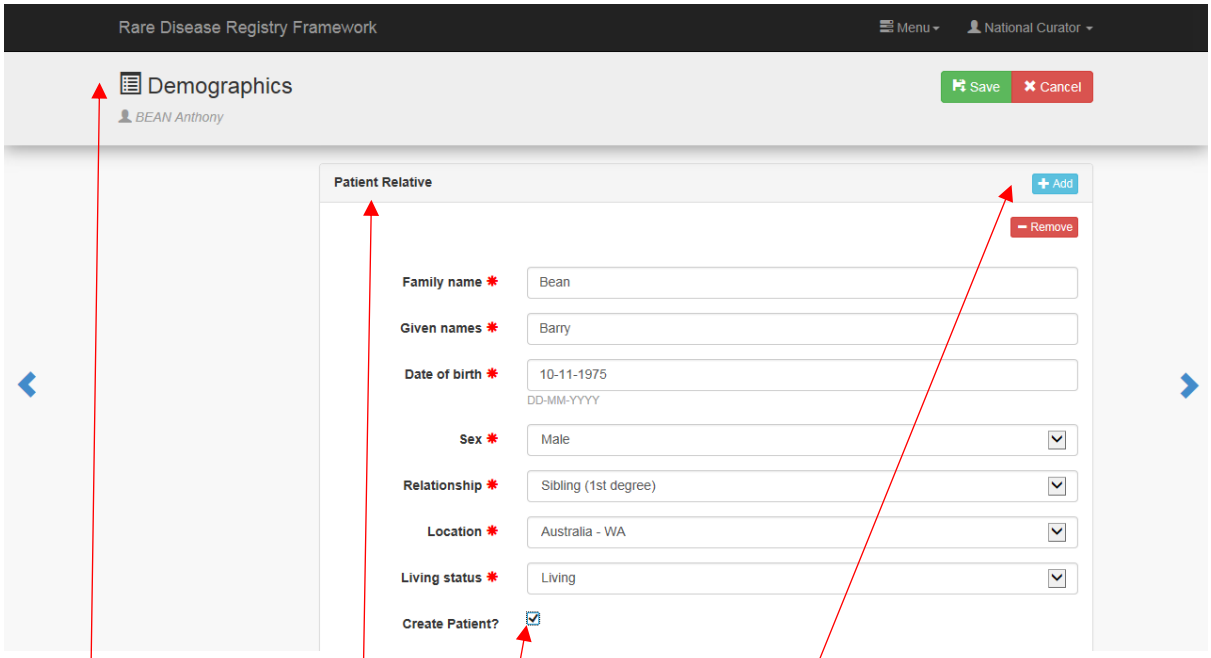
If you wish to delete the Doctor:

- Check the **Mark for deletion** box
- Click **Save**
- Doctor's details will be deleted

RETURN TO MENU

Add a Relative

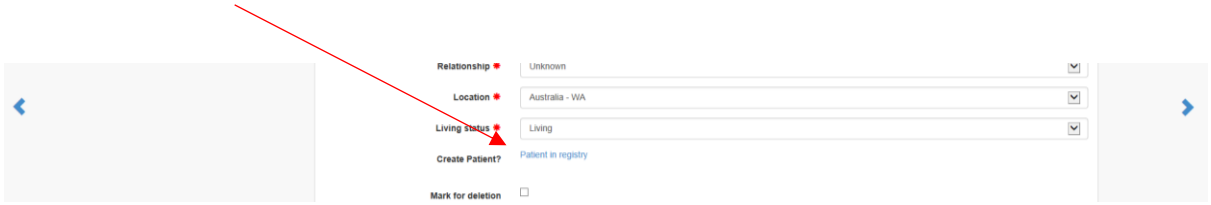
The Index patient's demographic page has an additional **Patient Relative** field, where all relatives are entered.



- Go to the Index's **Demographics** page
- Scroll down to **Patient Relative** field
- Click **+ Add** button and a new blank record will appear
- Complete the relative's details. All are required fields.
- Check **Create Patient?** tick box
- Click **Save** and set of modules will be created for this relative

The relative then can be accessed via the:

- Family Linkage or
- **Patient in registry** link which will appear after saving



RETURN TO MENU

Enter a Relative in a Different Centre to the Index

Relatives are automatically created with the same **Centre** as the Index.

If the index is not your patient you will not be able to access the index's demographics page to add your relative.

You will need to email the following details to the National Coodinator to arrange the relative to be added to the existing Index at another Centre.

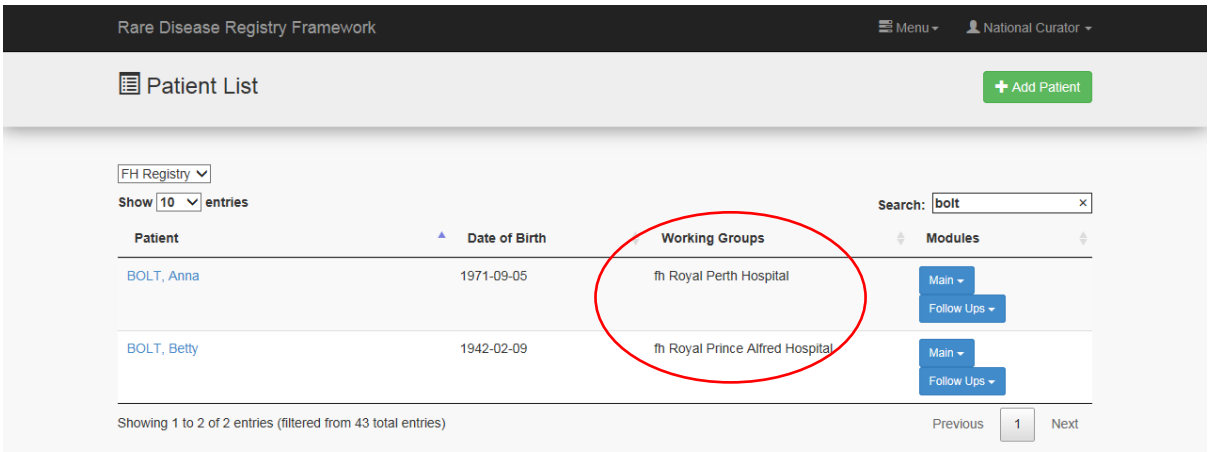
- Details of index
  - Family name
  - Given names
  - Date of birth
- Details of relative
  - Family name
  - Given names
  - Date of birth
  - Sex
  - Relationship (to the index)
  - Location
  - Living status

RETURN TO MENU

View Family Members in Different Centres

You will only be able to view patients in **Centres** you have permission to access, unless your patient has family members in other **Centres** and then they can be viewed via the Family Linkage function.

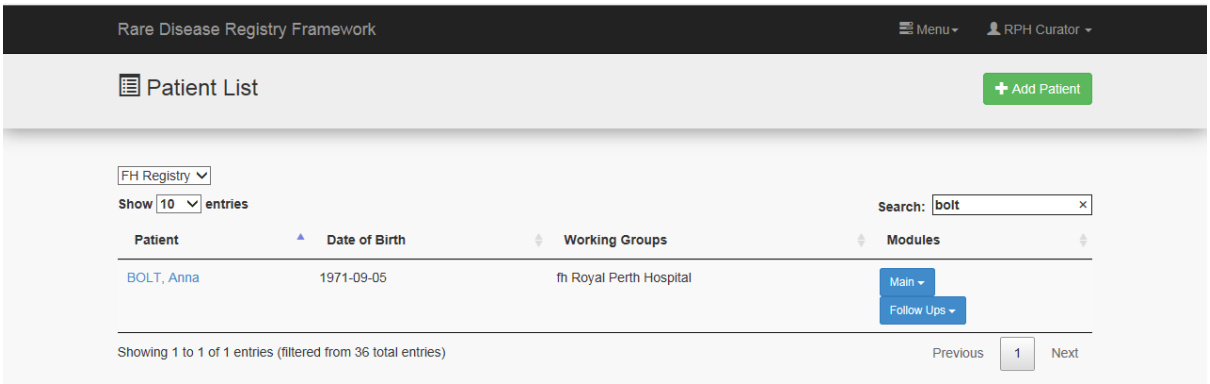
For example:



Anna Bolt (Index) is a patient at Royal Perth Hospital (RPH).

Her mother Betty Bolt (Relative) is a patient at Royal Prince Alfred Hospital (RPAH).

Only the National Coordinator can see this view.



The RPH coordinator can only see Anna Bolt (RPH patient) on their **Patient List** page.

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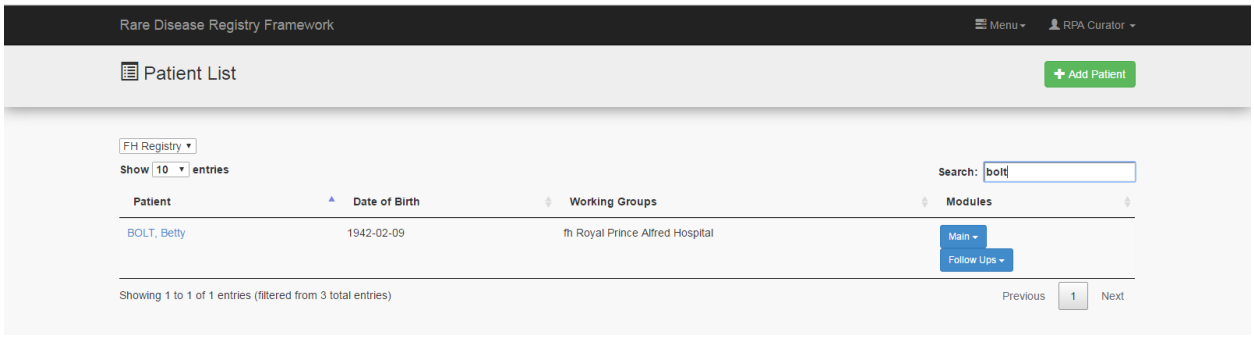
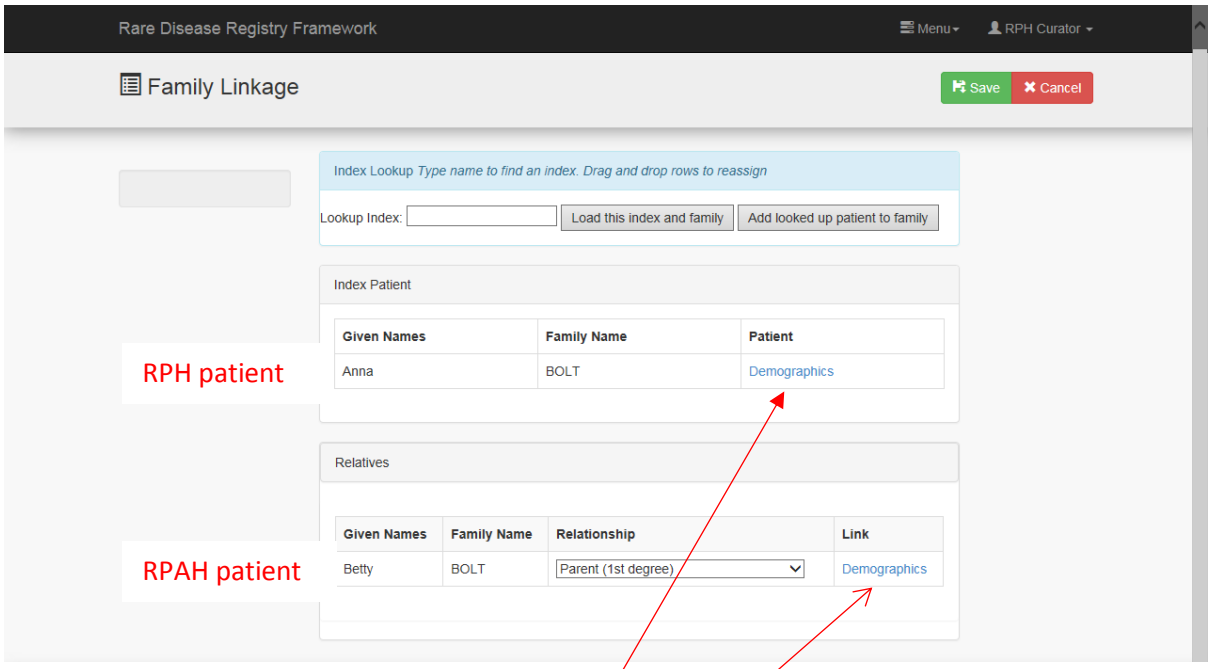


The RPAH coordinator can only see Betty Bolt (RPAH patient) on their **Patient List** page.

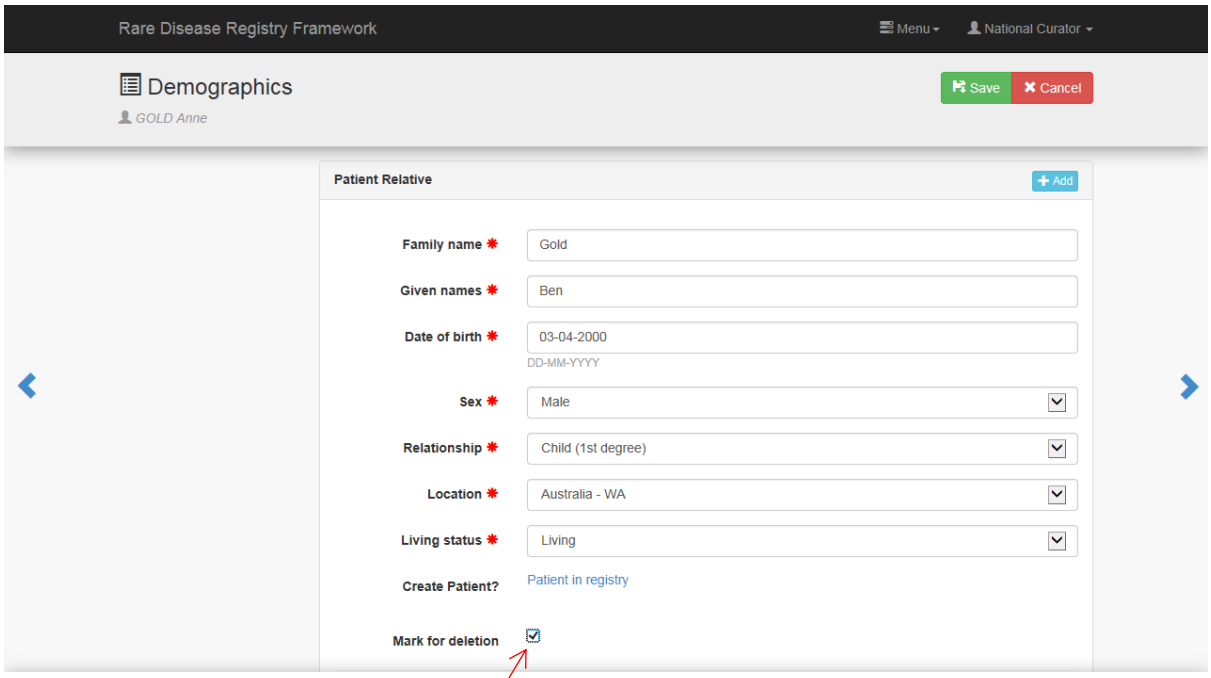


On the **Family Linkage** page:

- The RPAH coordinator can click the **Demographics** and access Anna Bolt's (RPH patient) results
  N.B. Anna Bolt's results will be a 'view only' access for the RPAH coordinator.
- The RPH coordinator can click the **Demographics** and access Betty Bolt's (RPAH patient) results
  N.B. Betty Bolt's results will be a 'view only' access for the RPH coordinator.

RETURN TO MENU

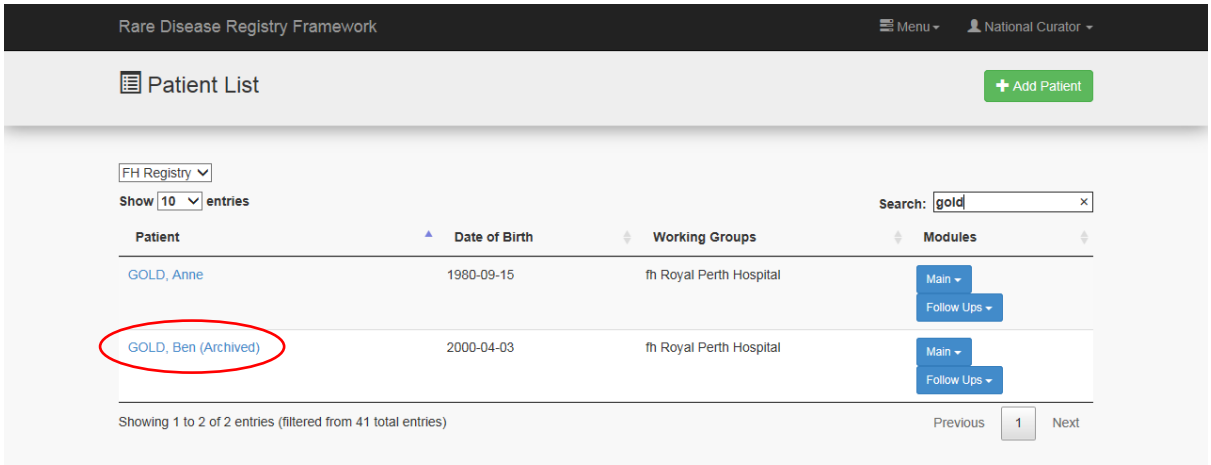
Delete a Relative



Only relatives can be deleted directly. To delete an Index, the Index must be changed to a relative (see Change the Index).

To delete a relative:

- Go to the Index's Demographics page
- Scroll down to the **Patient Relative** section
- Check the **Mark for deletion** box on the relative you wish to delete
- Click **Save**



The deleted patient is now shown as **(Archived)** on the **Patient List** page.

The deleted patient will no longer appear on the **Family Linkage** or in the **Patient Relative** field.

All archived patients will be deleted periodically by the database administrator.

RETURN TO MENU

Delete an Index

Only relatives can be deleted directly.

To delete an Index, the Index must first be changed to a Relative (Change the Index) and then deleted (Delete a Relative).

To delete an Index with at least 1 relative:

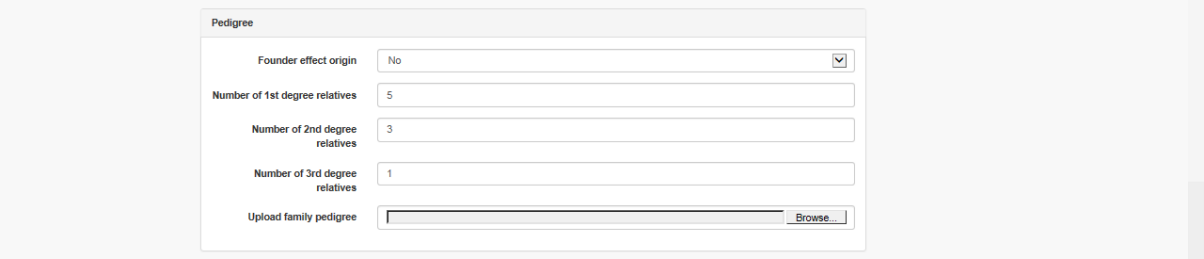
- Use the Change the Index function (makes the Index a Relative)
- Use the Delete a Relative function

To delete an Index with no relatives:

- Change the **Family name** to 'DELETE'
- Change the **Given name** to 'DELETE'
- Email details to the National Coordinator
- Database administrator will delete the patient

RETURN TO MENU

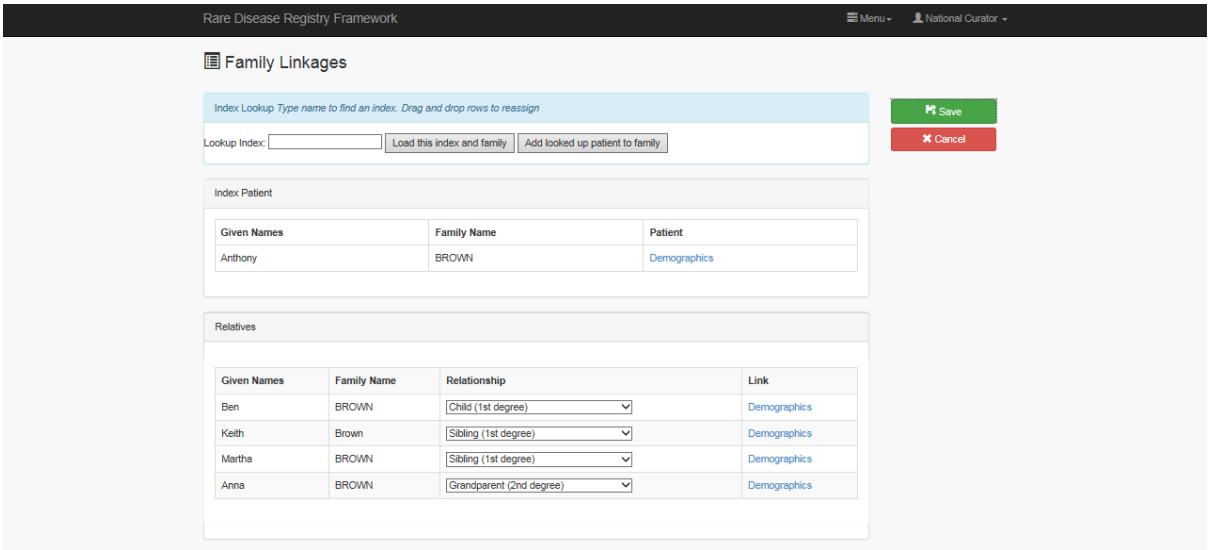
Pedigree



The pedigree indicates the number of possible relatives.

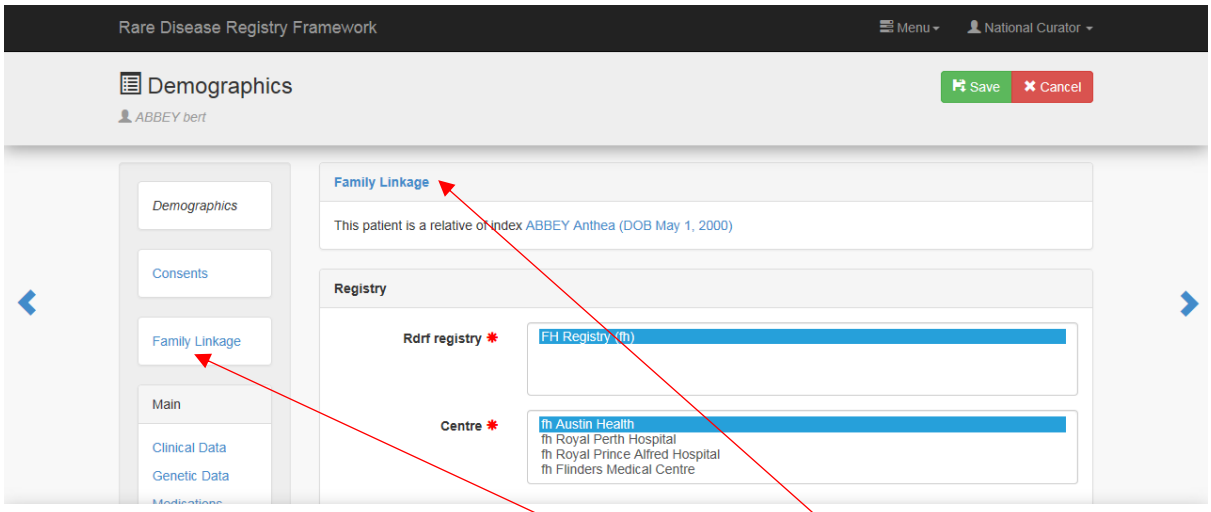
When compared with the **Family Linkage** (the number of family members already in the Registry) you can see:

- For 1st degree relatives you have consented 3 out of the possible 5 relatives
- For 2nd degree relatives you have consented 1 out of the possible 3 relatives
- For 3rd degree relatives you have consented 0 out of the possible 1 relative

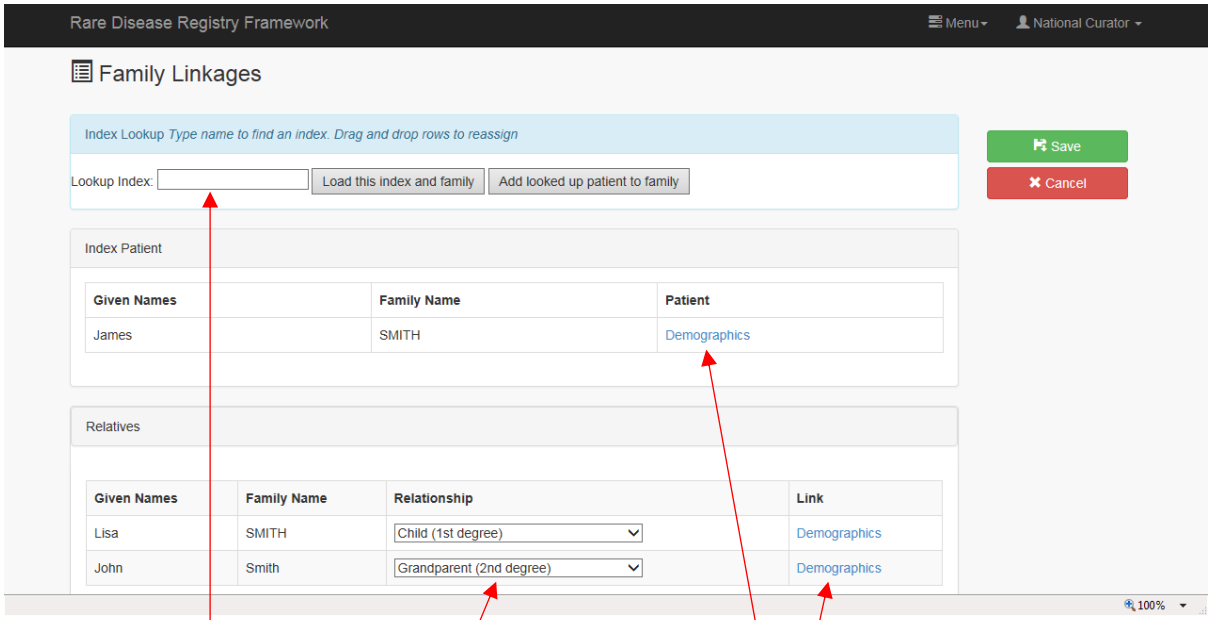


RETURN TO MENU

Family Linkage (screen shot – Family Linkage)



The **Family Linkage** can be accessed via the menu on the left or the **Family Linkage** link.

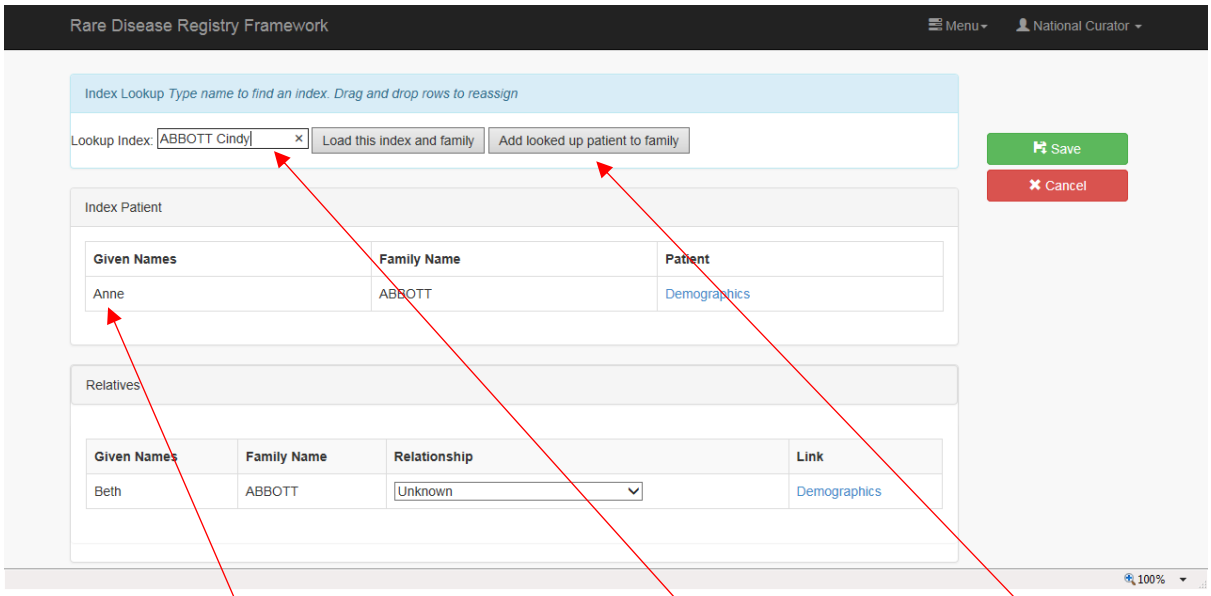


On the **Family Linkage** screen you can:

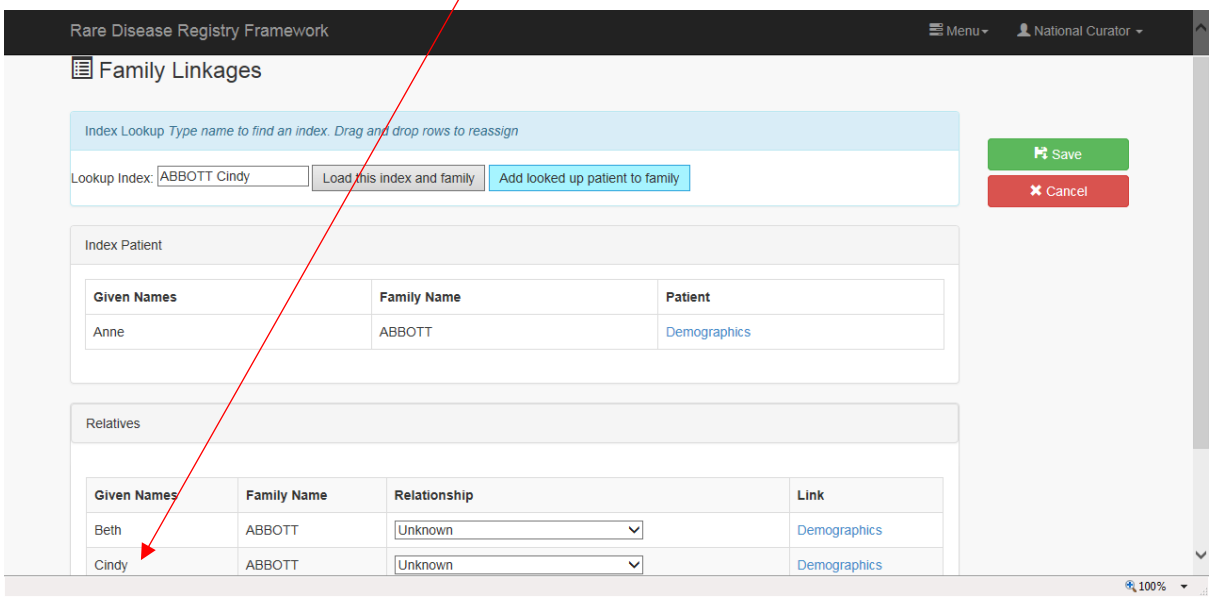
- Move between family members by clicking on the **Demographics** links
- See family relationships
- Load another Index and Relatives by typing in the name of the Index in the **Lookup index** field and clicking **Load this index and family** button
- Change an incorrectly entered Index patient to a Relative
- Change the Index

RETURN TO MENU

Change an incorrectly entered Index patient to a Relative



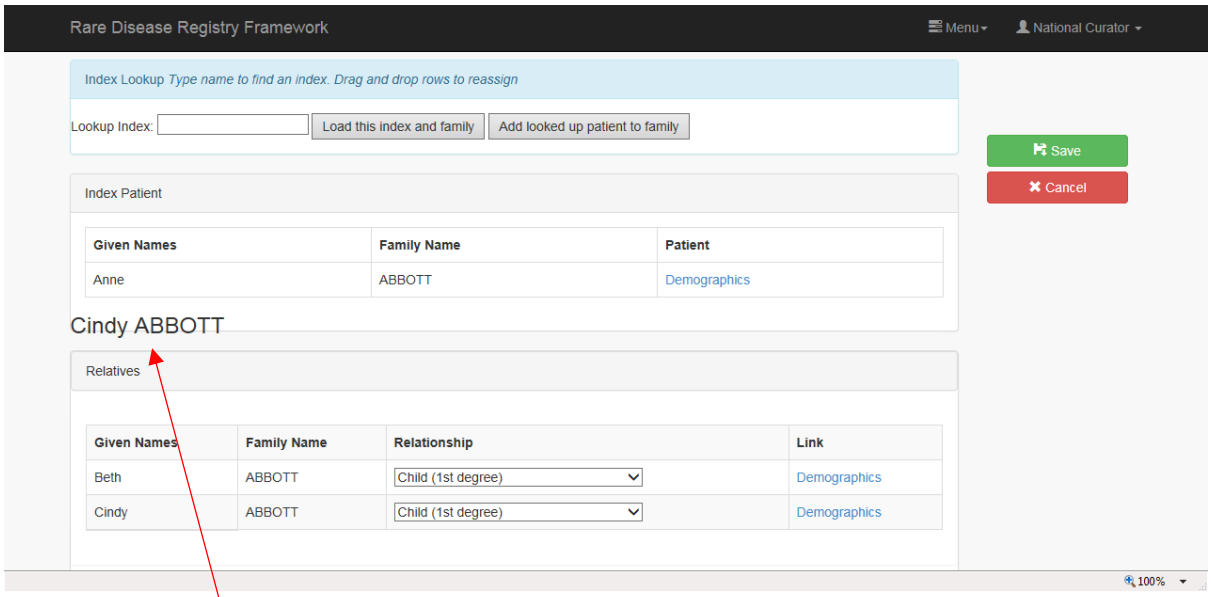
- Load the family you wish to add the incorrect Index patient to. This can be done by going to any member of the family and clicking on the **Family Linkage.**
- Typing in the name of the incorrect Index in the **Lookup index** field and click **Add looked up patient to family** button
- The incorrect **Index** will now be a **Relative**
- Enter **Relationship**
- Click **Save**



- Go to **Patient Relative** field on Index's **Demographic** page and complete missing details on the added relative
- Click **Save**

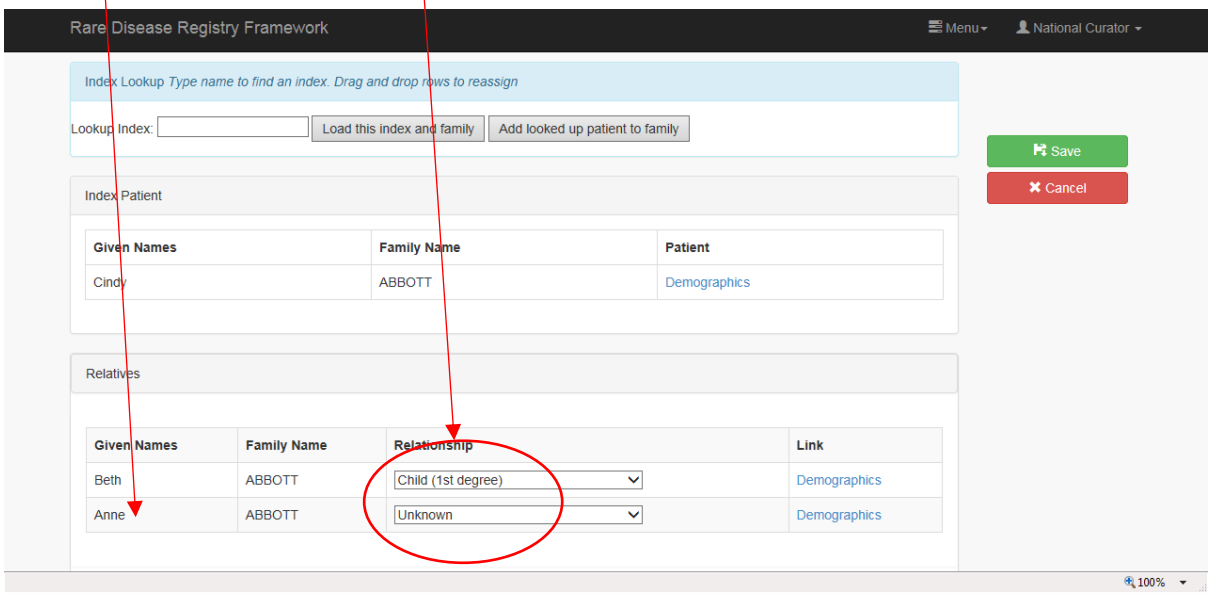
RETURN TO MENU

Change the Index



- Go to the **Family Linkage** page
- Drag the **Relative** you wish to become the new index to the **Index Patient** field
- Click **Save**
- The old index is now a relative and can now be deleted from the Registry if required. See Delete a Relative.

Warning: Changing the Index should be avoided and only used in rare cases of the Index withdrawing from the Registry. Note the **Relationships** will now be incorrect and will need to be re-entered. Multiple fields in the **Patient Relative** section will also be lost and need to be re-entered.



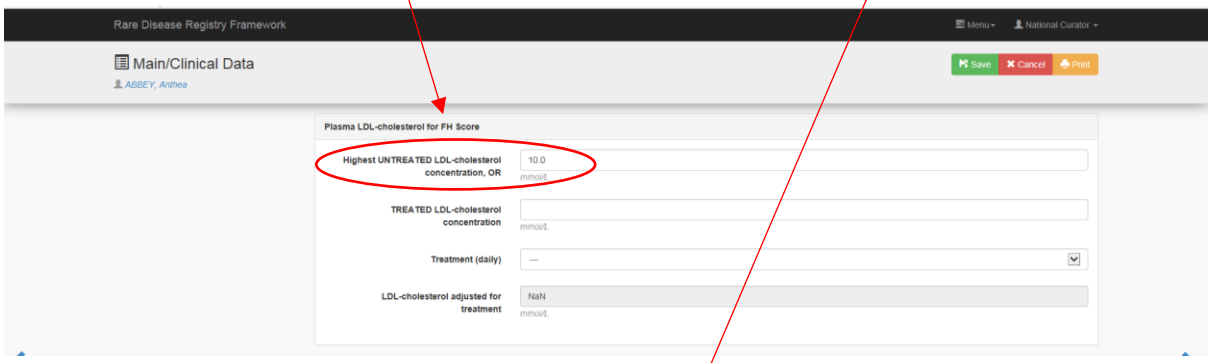
RETURN TO MENU

Clinical Data (screen shot – Clinic Data)

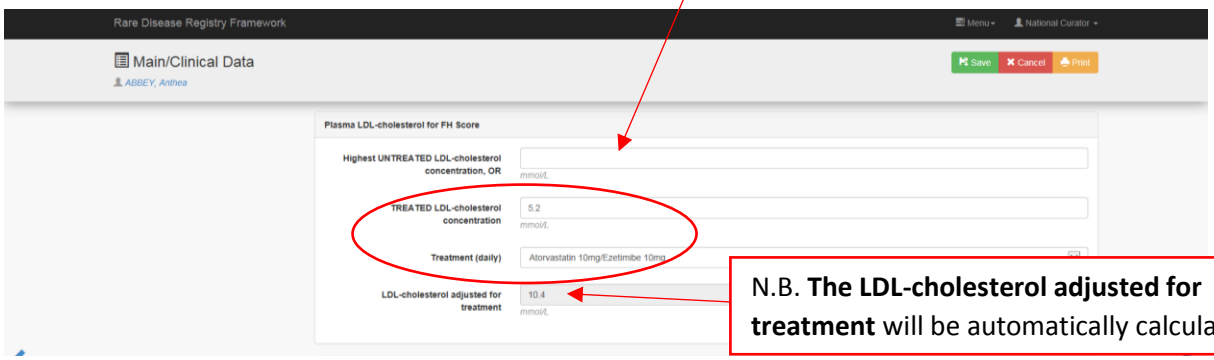
Calculate the DLCNS

To calculate the DLCNS:

- Complete all the required fields (marked with an *) including the **Date of assessment**
- Complete the **Plasma LDL-cholesterol for FH Score** section. **Complete either:**
  - Highest UNTREATED LDL-cholesterol concentration (on record)
    OR
  - TREATED LDL-cholesterol concentration AND Treatment (daily)



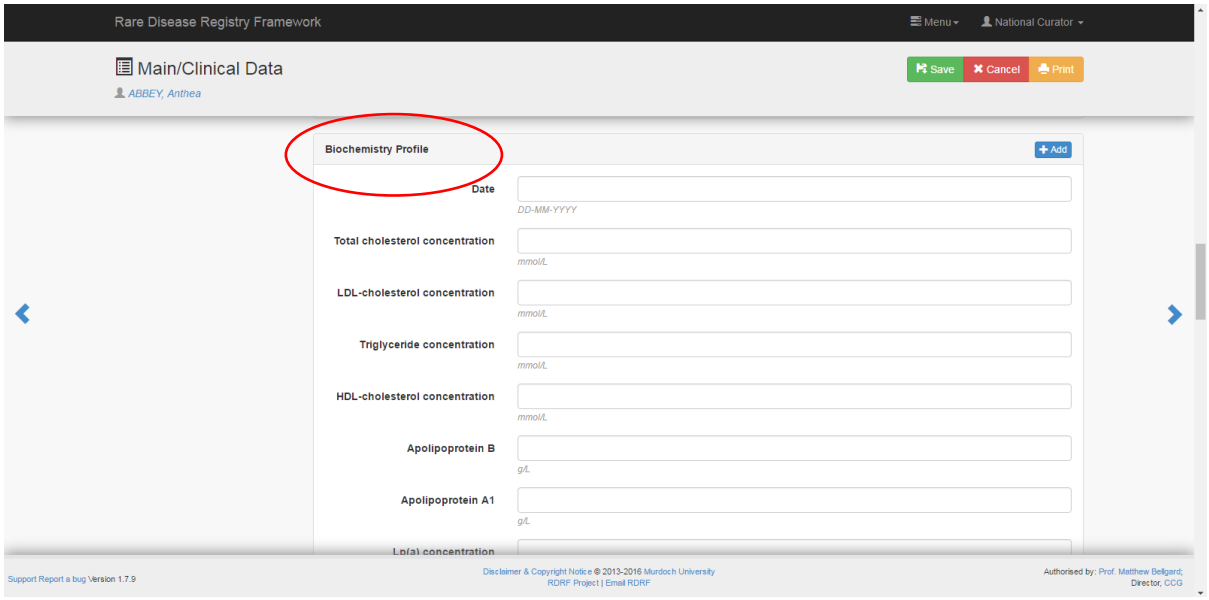
OR



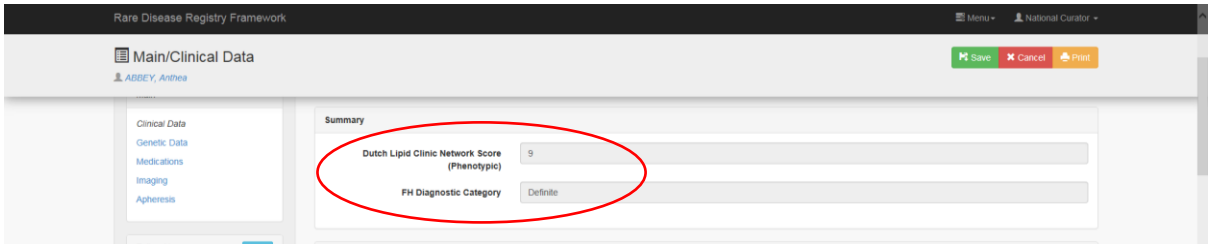
- Click **Save**

N.B. If both the UNTREATED and TREATED LDL-cholesterol are entered the **UNTREATED LDL-cholesterol** will be used for the calculation of the DLCNS.

RETURN TO MENU



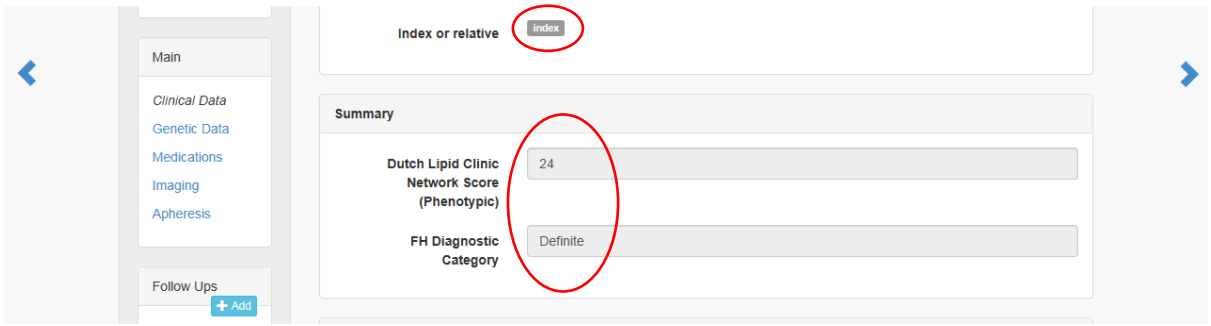
N.B. Enter current lipid profile in the **Biochemistry Profile** section



Depending on the type/age of the patient (see Adult Index, Child/adolescent Index or Relative) a **Dutch Lipid Clinic Network Score (Phenotype)** and/or **FH Diagnostic Category** will automatically appear in the **Summary** section.

Adult Index

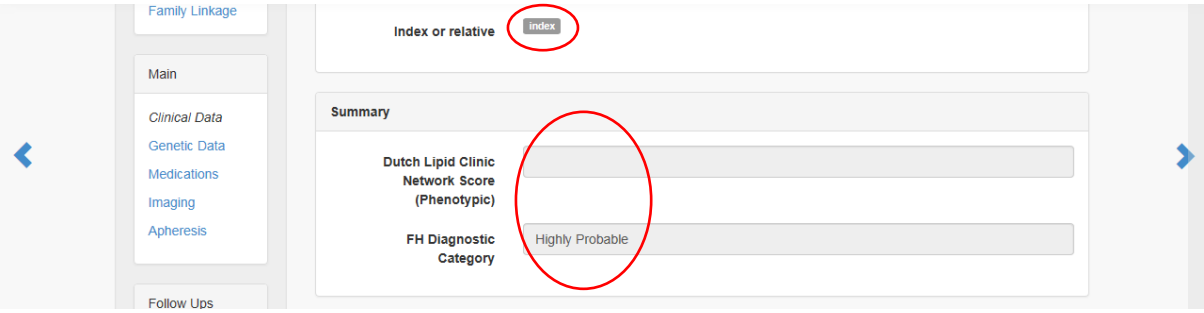
Adult INDEX will be given a **DLCNS** and an **FH Diagnostic Category**.



RETURN TO MENU

Child/adolescent Index

Child/adolescent INDEX will be given an **FH Diagnostic Category** only.



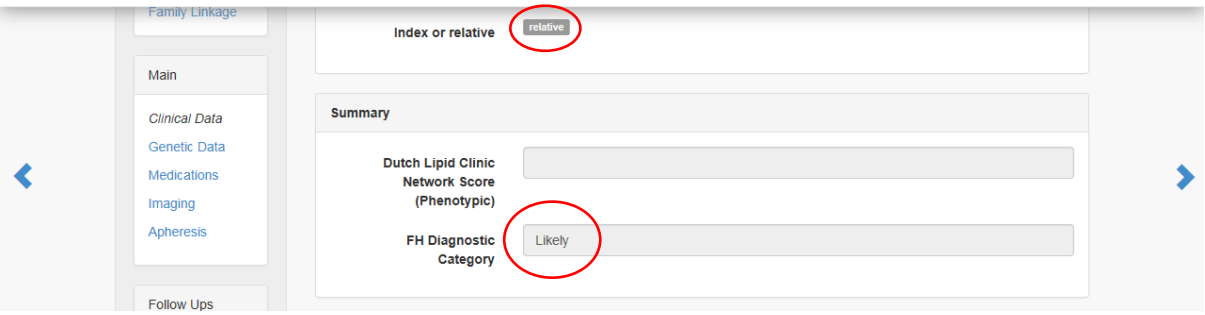
To be included in the registry as an INDEX, a child/adolescent must have a **highly probable** or **probable** category.

Category	Inclusion as an INDEX	Criteria
Highly probable	Yes	LDL-cholesterol concentration > 5.0 mmol/L
Probable	Yes	• LDL-cholesterol concentration > 4.0 - 5.0 mmol/L + family history of premature CHD or • LDL-cholesterol concentration > 4.0 - 5.0 mmol/L + hypercholesterolaemia or tendon xanthoma or arcus in first-degree relative
Possible	No	LDL-cholesterol concentration 4.0 - 5.0 mmol/L
Unlikely	No	LDL-cholesterol concentration < 4.0 mmol/L

RETURN TO MENU

Relative

All relatives including children and adolescents will be given an **FH Diagnostic Category** only, based on their LDL-cholesterol level for their age and gender (see tables below).



Likely - red
Uncertain - grey
Unlikely – green

LDL-C FEMALES

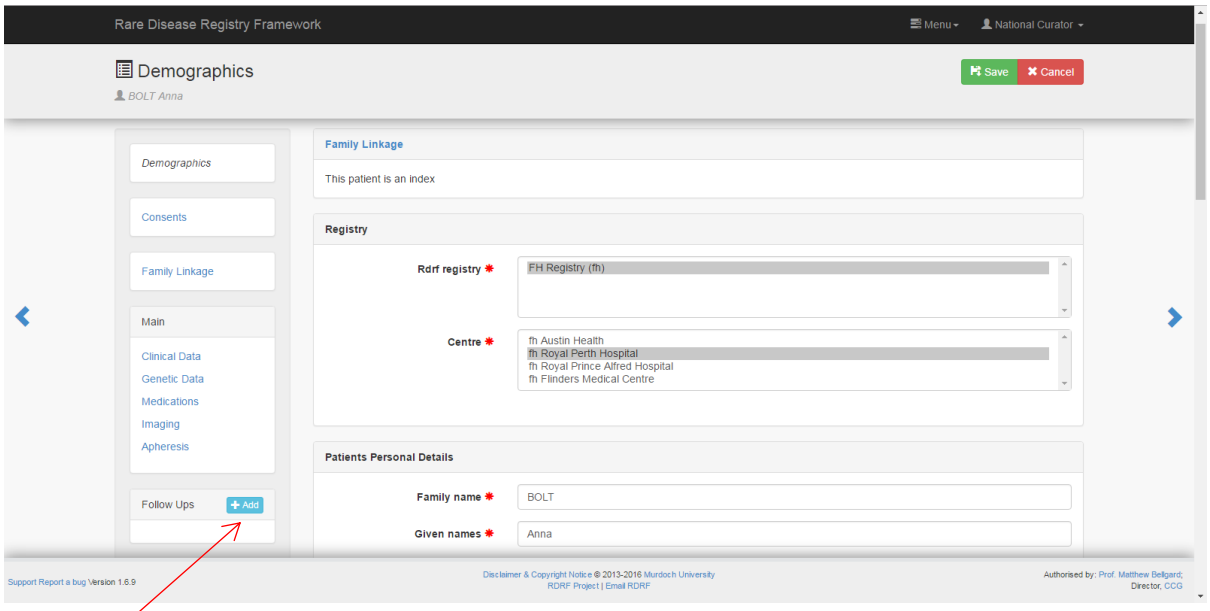
Age					
0 To 14	15 To 24	25 To 34	35 To 44	45 To 54	55 and Older
5.5	5.5	5.5	5.5	5.5	5.5
5.4	5.4	5.4	5.4	5.4	5.4
5.3	5.3	5.3	5.3	5.3	5.3
5.2	5.2	5.2	5.2	5.2	5.2
5.1	5.1	5.1	5.1	5.1	5.1
5.0	5.0	5.0	5.0	5.0	5.0
4.9	4.9	4.9	4.9	4.9	4.9
4.8	4.8	4.8	4.8	4.8	4.8
4.7	4.7	4.7	4.7	4.7	4.7
4.6	4.6	4.6	4.6	4.6	4.6
4.5	4.5	4.5	4.5	4.5	4.5
4.4	4.4	4.4	4.4	4.4	4.4
4.3	4.3	4.3	4.3	4.3	4.3
4.2	4.2	4.2	4.2	4.2	4.2
4.1	4.1	4.1	4.1	4.1	4.1
4.0	4.0	4.0	4.0	4.0	4.0
3.9	3.9	3.9	3.9	3.9	3.9
3.8	3.8	3.8	3.8	3.8	3.8
3.7	3.7	3.7	3.7	3.7	3.7
3.6	3.6	3.6	3.6	3.6	3.6
3.5	3.5	3.5	3.5	3.5	3.5
3.4	3.4	3.4	3.4	3.4	3.4
3.3	3.3	3.3	3.3	3.3	3.3
3.2	3.2	3.2	3.2	3.2	3.2
3.1	3.1	3.1	3.1	3.1	3.1
3.0	3.0	3.0	3.0	3.0	3.0

LDL-C MALES

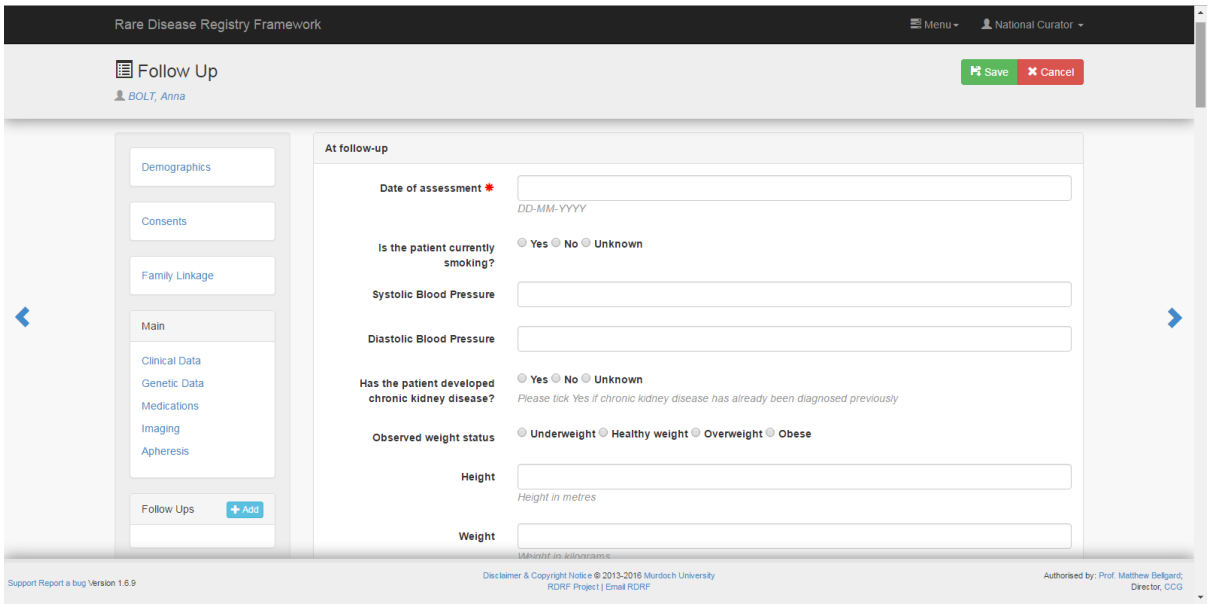
Age					
0 To 14	15 To 24	25 To 34	35 To 44	45 To 54	55 and Older
5.5	5.5	5.5	5.5	5.5	5.5
5.4	5.4	5.4	5.4	5.4	5.4
5.3	5.3	5.3	5.3	5.3	5.3
5.2	5.2	5.2	5.2	5.2	5.2
5.1	5.1	5.1	5.1	5.1	5.1
5.0	5.0	5.0	5.0	5.0	5.0
4.9	4.9	4.9	4.9	4.9	4.9
4.8	4.8	4.8	4.8	4.8	4.8
4.7	4.7	4.7	4.7	4.7	4.7
4.6	4.6	4.6	4.6	4.6	4.6
4.5	4.5	4.5	4.5	4.5	4.5
4.4	4.4	4.4	4.4	4.4	4.4
4.3	4.3	4.3	4.3	4.3	4.3
4.2	4.2	4.2	4.2	4.2	4.2
4.1	4.1	4.1	4.1	4.1	4.1
4.0	4.0	4.0	4.0	4.0	4.0
3.9	3.9	3.9	3.9	3.9	3.9
3.8	3.8	3.8	3.8	3.8	3.8
3.7	3.7	3.7	3.7	3.7	3.7
3.6	3.6	3.6	3.6	3.6	3.6
3.5	3.5	3.5	3.5	3.5	3.5
3.4	3.4	3.4	3.4	3.4	3.4
3.3	3.3	3.3	3.3	3.3	3.3
3.2	3.2	3.2	3.2	3.2	3.2
3.1	3.1	3.1	3.1	3.1	3.1
3.0	3.0	3.0	3.0	3.0	3.0

RETURN TO MENU

Add Follow Up Form



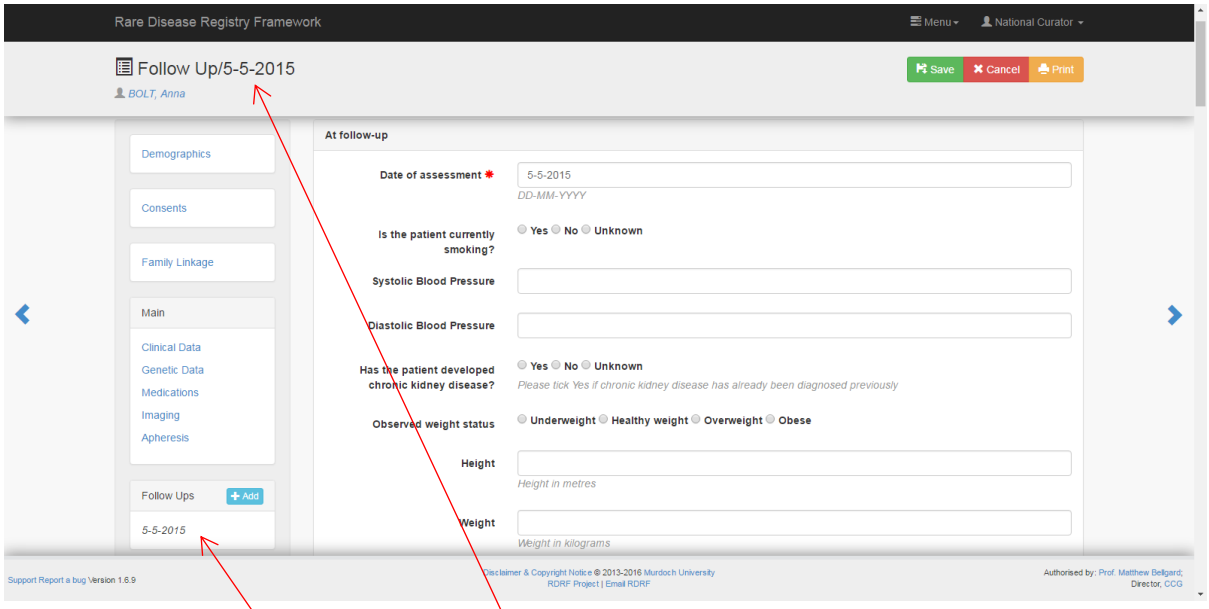
Click **+Add** button



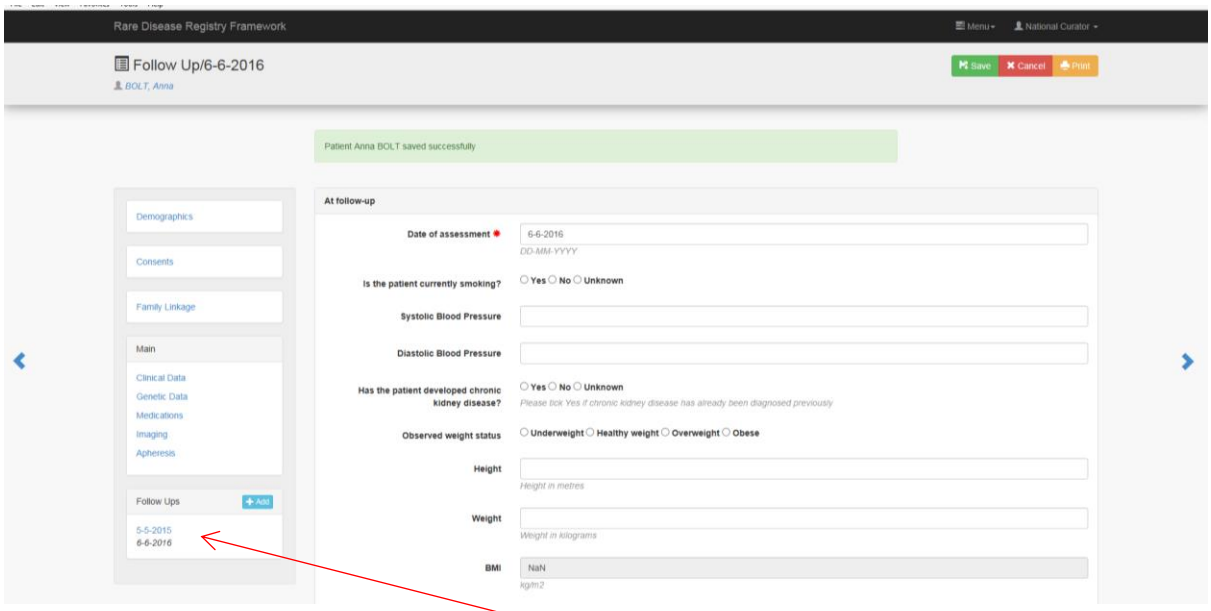
Blank **Follow Up** page will appear:

- Complete **Date of assessment** (required field)
- Complete other data
- Click **Save**

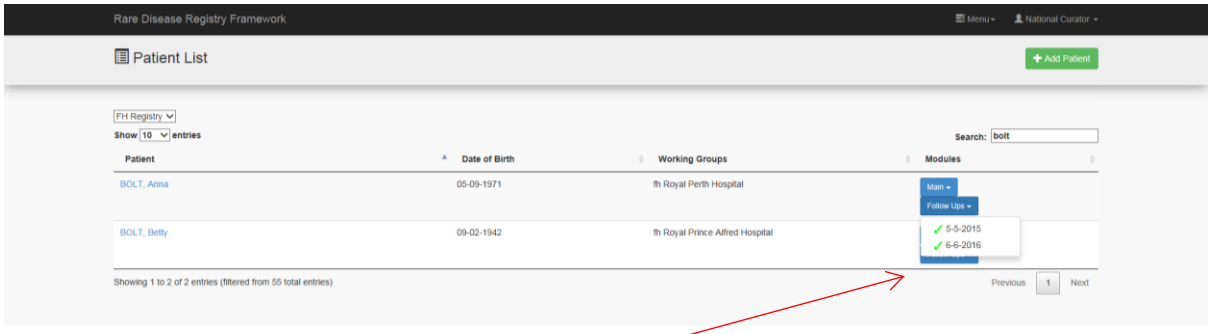
RETURN TO MENU



Date appears in left hand menu bar and header



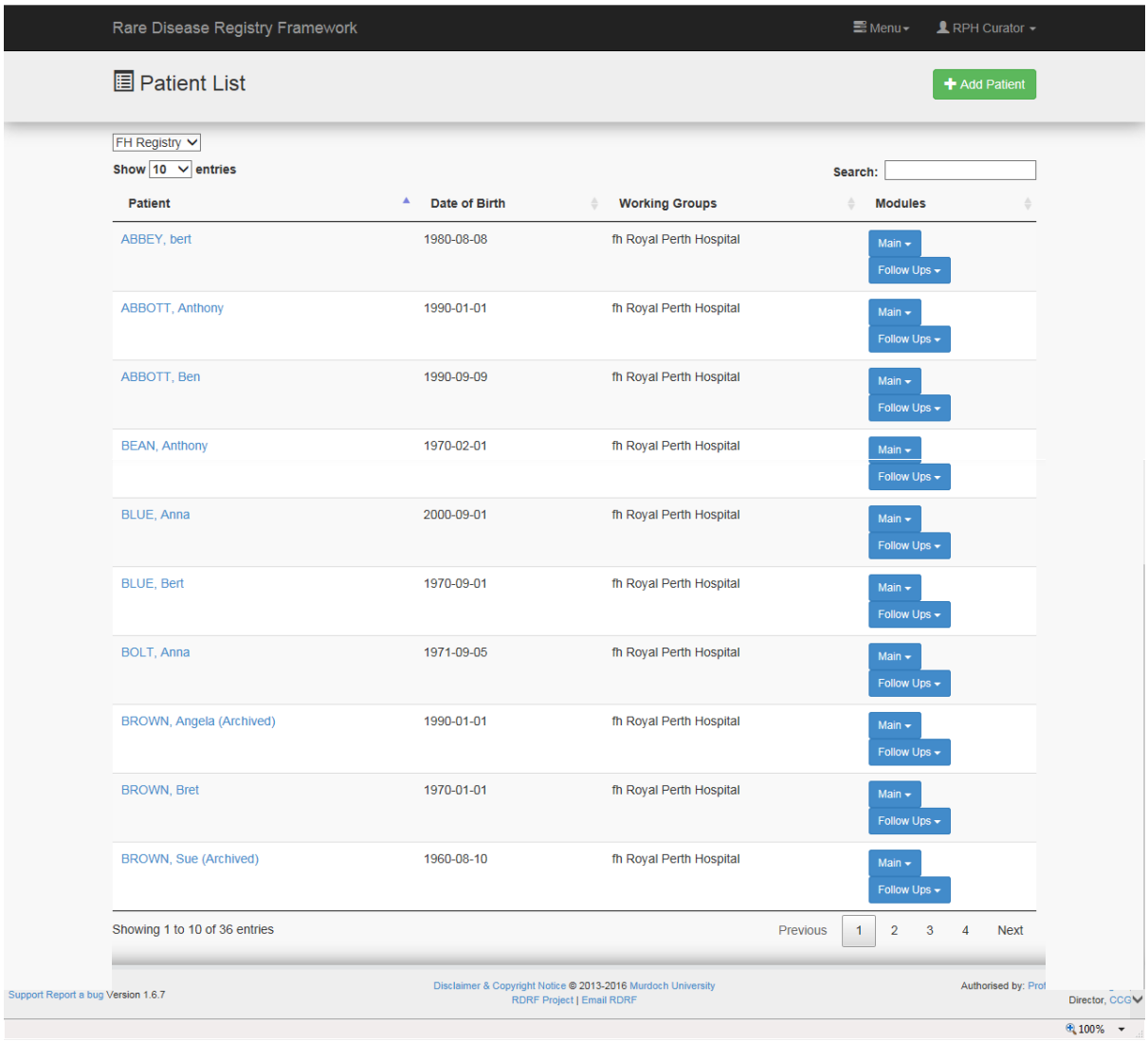
With multiple forms dates will appear in chronological order



Patient List page will show **Follow Ups** in dropdown menu

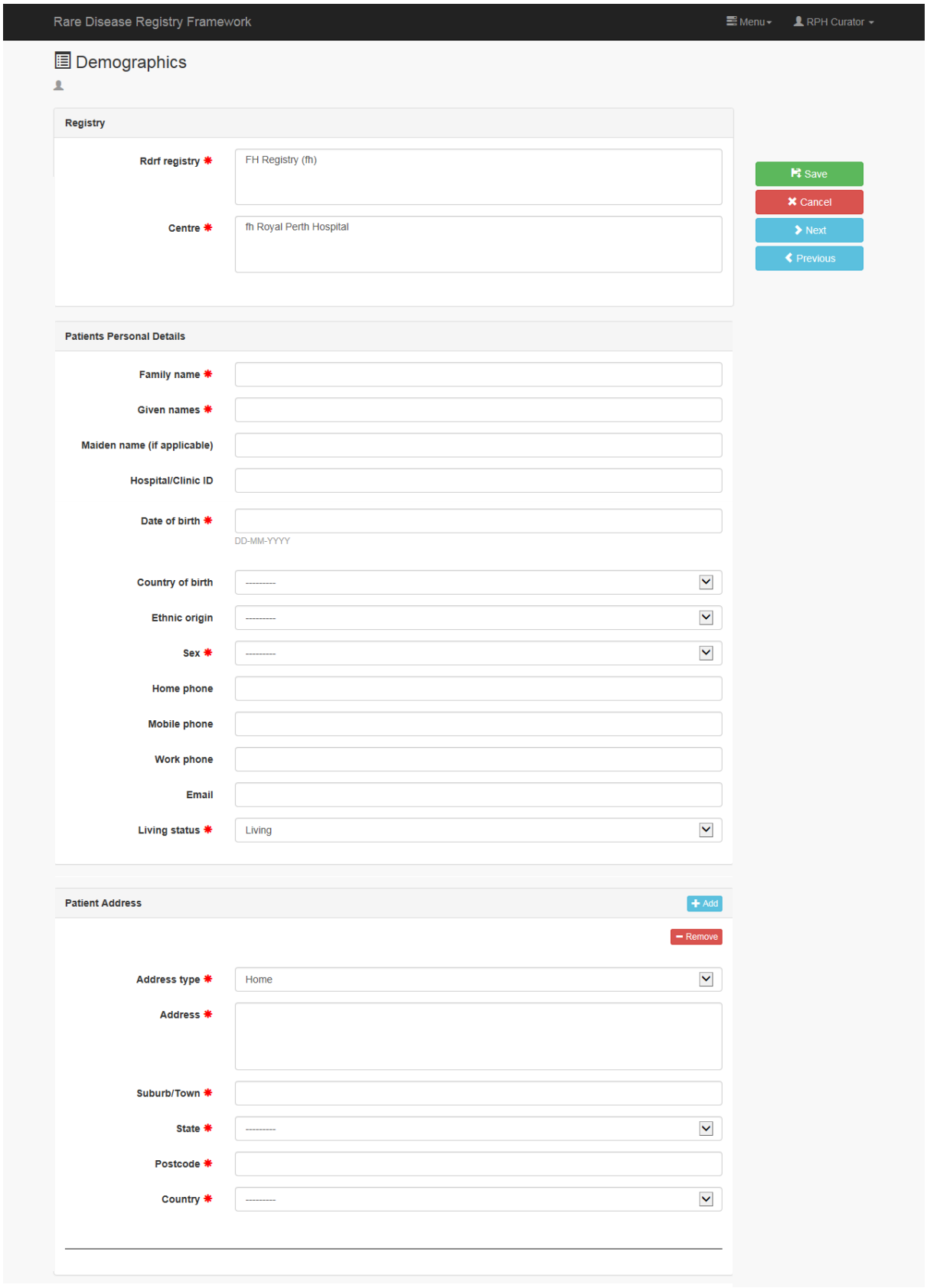
RETURN TO MENU

Screen Shot – Patient List (enlarge to view)

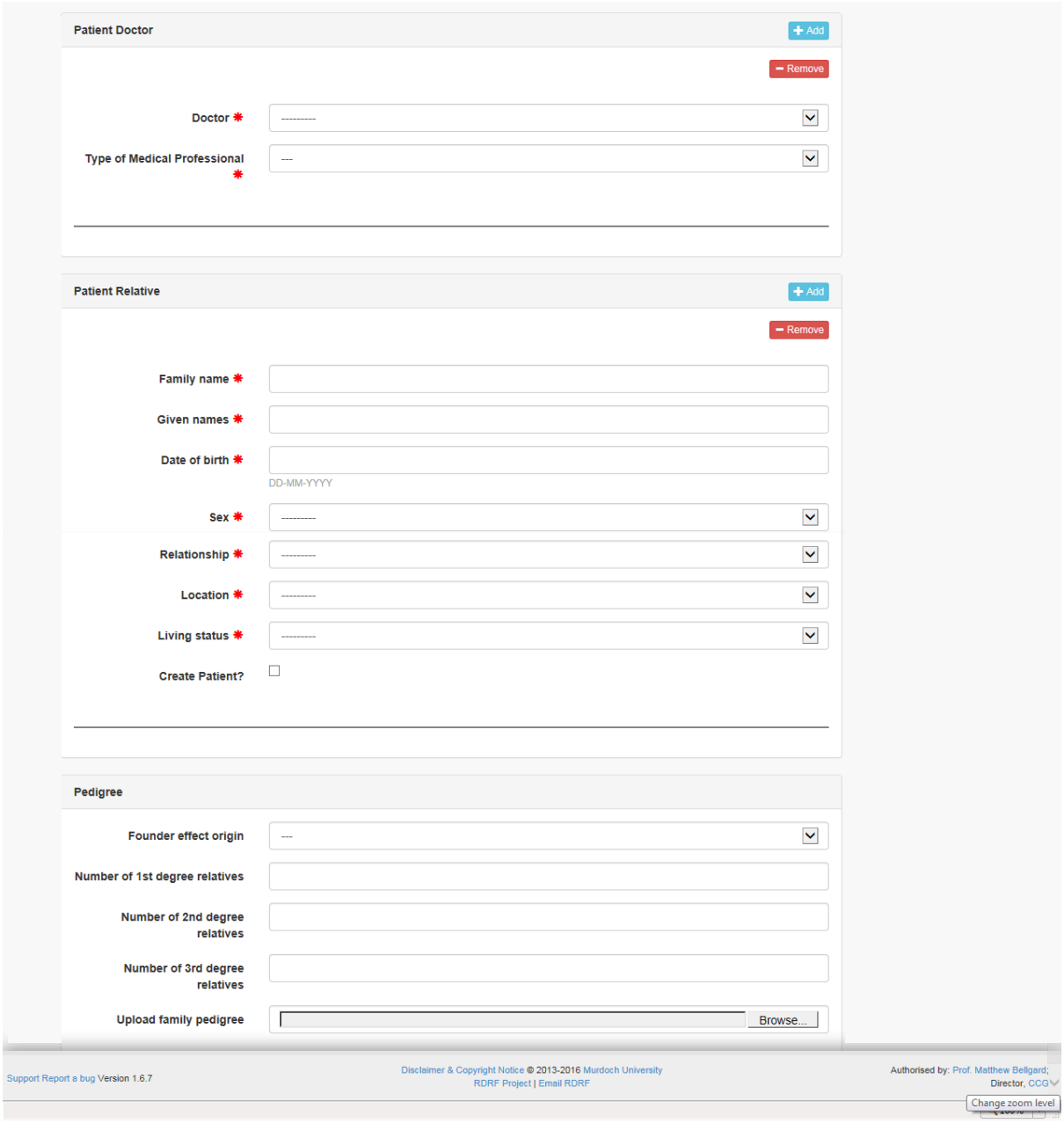


RETURN TO MENU

Screen Shot – Demographics (enlarge to view)

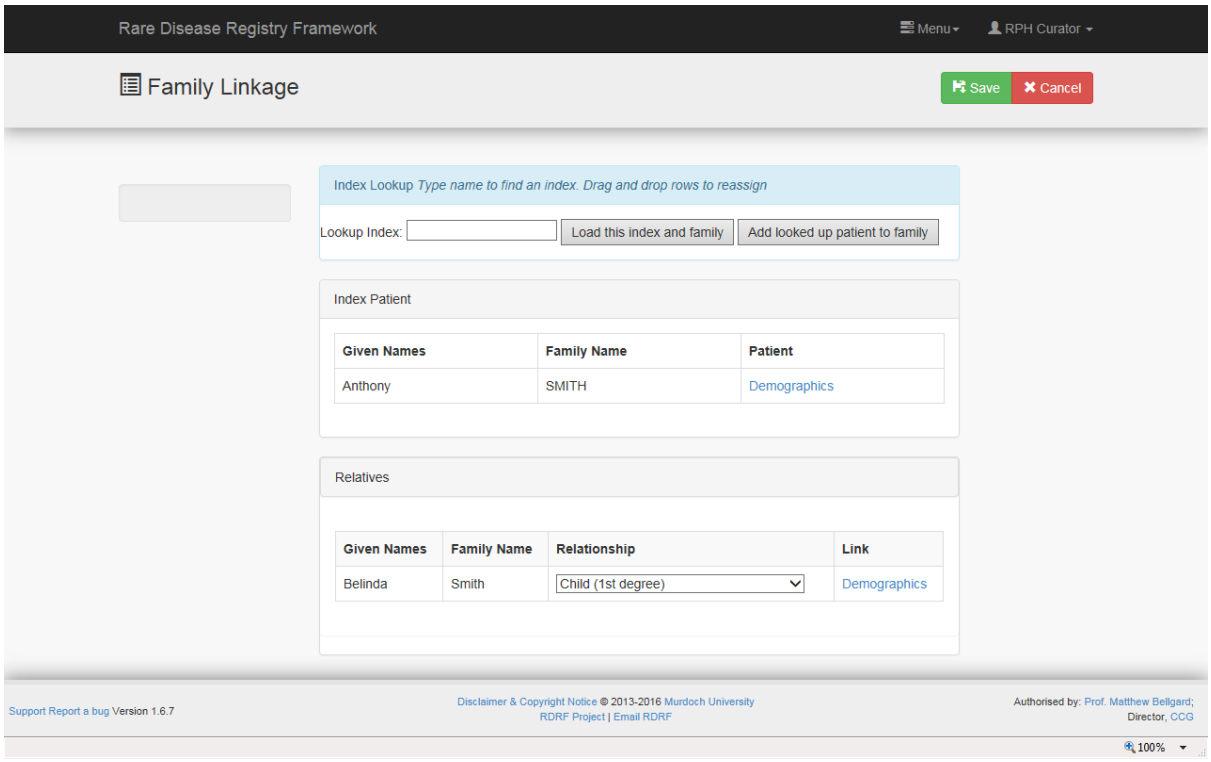


RETURN TO MENU



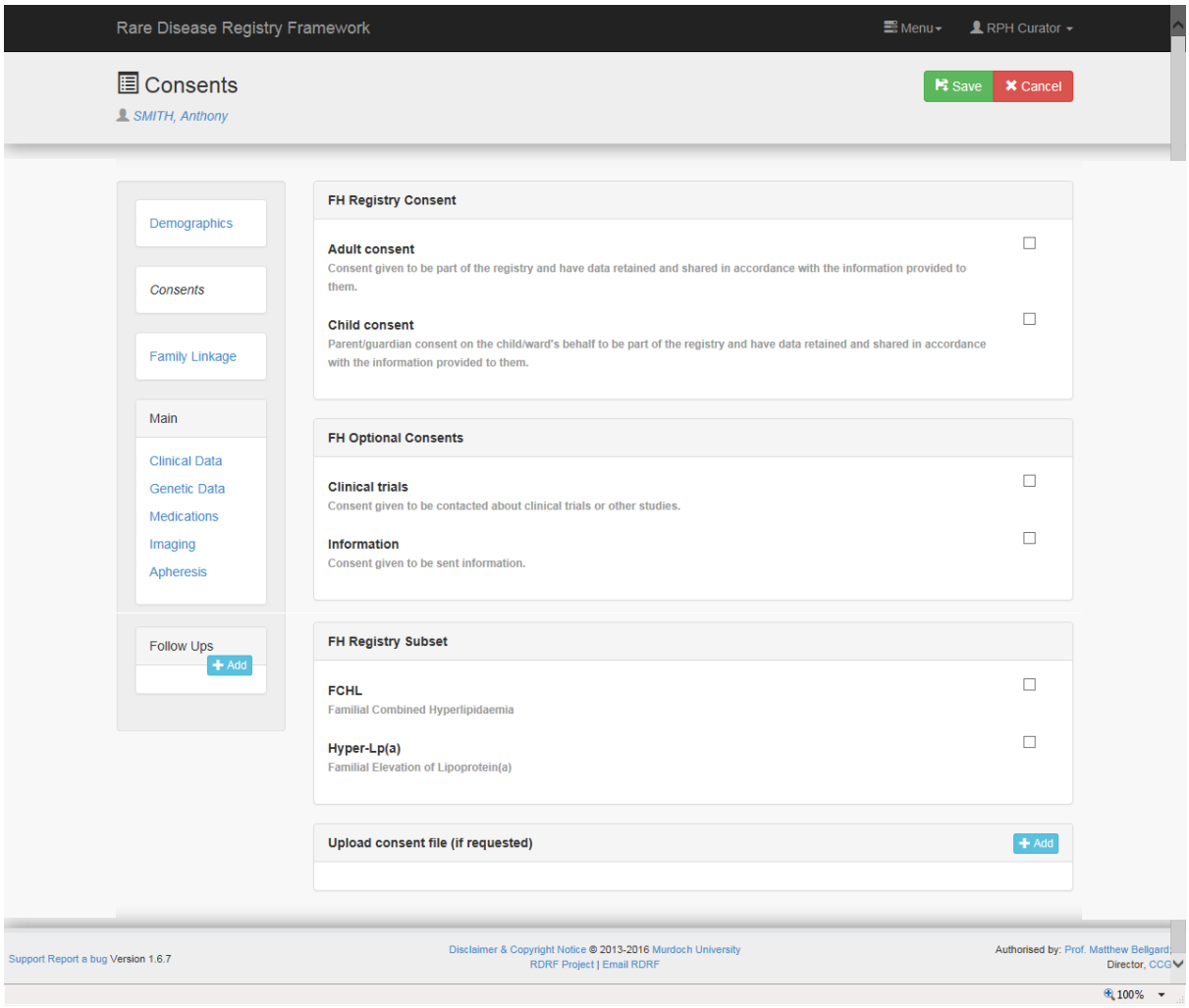
RETURN TO MENU

Screen Shot – Family Linkage (enlarge to view)



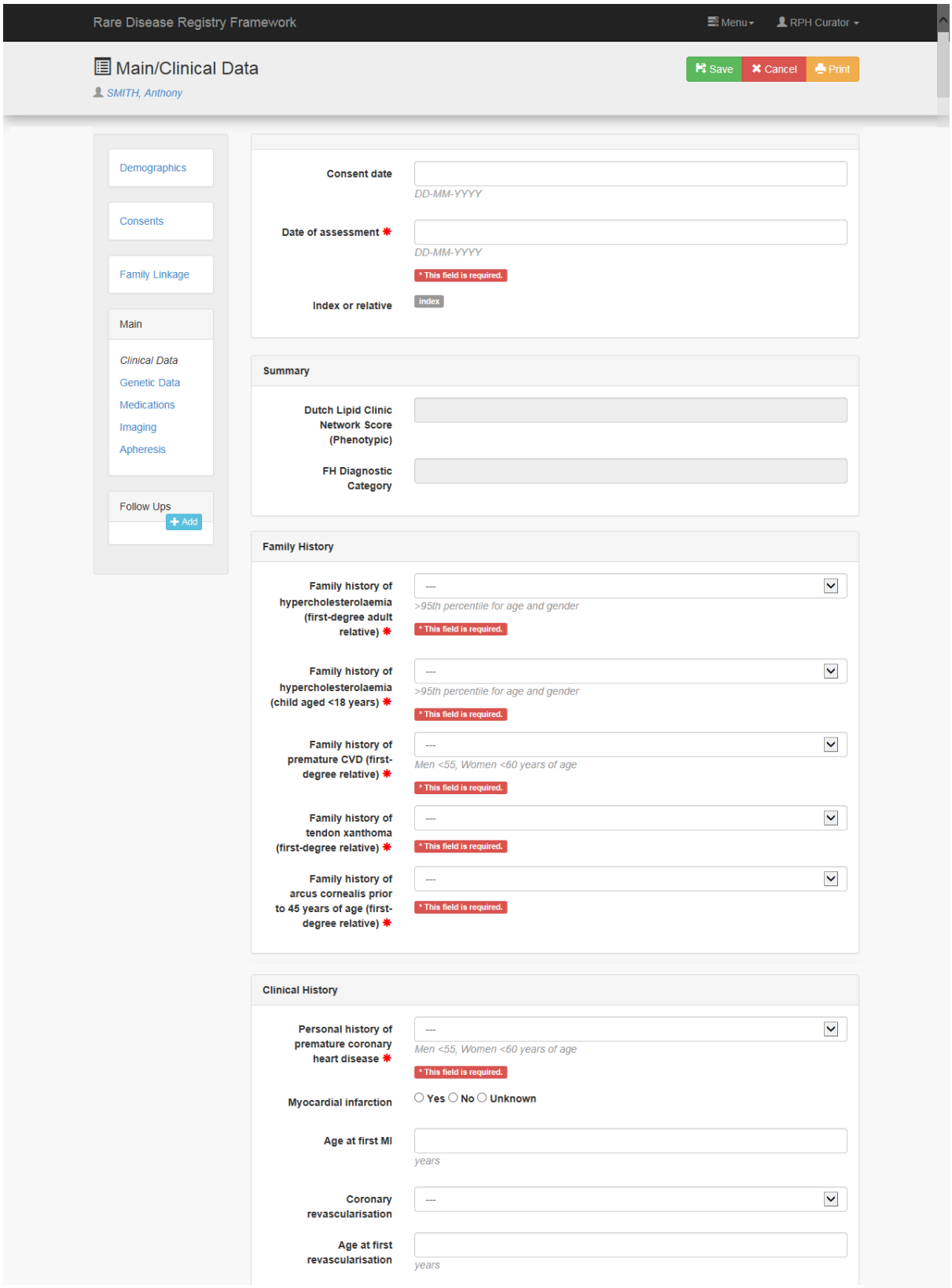
RETURN TO MENU

Screen Shot – Consents (enlarge to view)

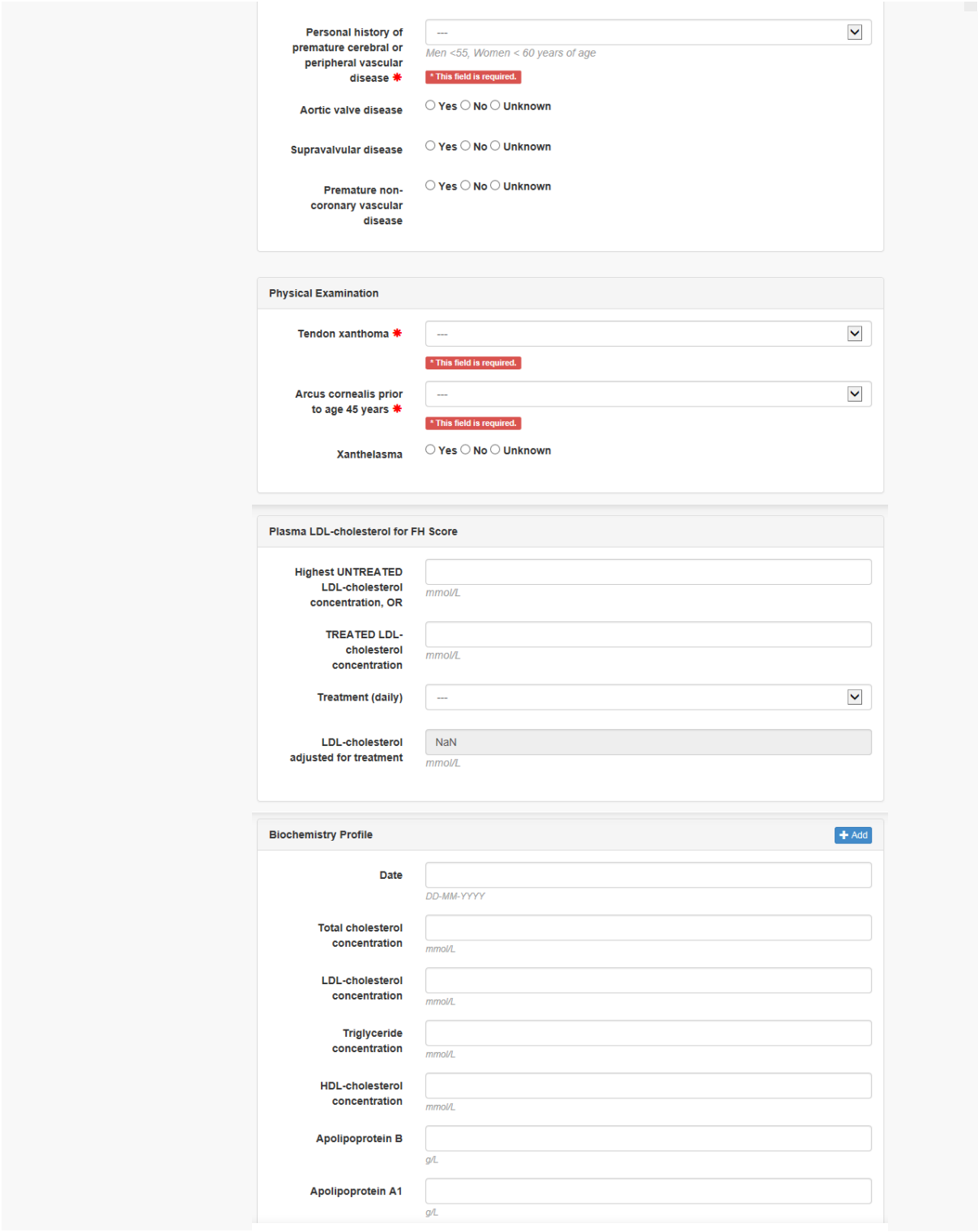


RETURN TO MENU

Screen Shot – Clinical Data (enlarge to view)



RETURN TO MENU



RETURN TO MENU

Lp(a) concentration
g/L

Aspartate transaminase (AST)
U/L

Alanine transaminase (ALT)
U/L

Creatine kinase (CK)
U/L

Albumin
g/L

Creatinine
umol/L

C-reactive protein (CRP)
mg/L

Treatment (daily) ---

If other, enter medication name(s), dose(s) and regime

Compliance ---
If intolerant, please enter details into Medication Module - Drug Intolerance

Mark for deletion ☐

Other CVD Risk Factors

Smoking ○ Yes ○ Never ○ Ex-smoker

Indicate pack years smoked
Pack years = (number of cigarettes smoked per day/20) x number of years smoked

Alcohol ---

Hypertriglyceridaemia ○ Yes ○ No ○ Unknown

Hypertension ---
If yes and on hypertensive medication, please complete the Medication Module

Systolic Blood Pressure

Diastolic Blood Pressure

Heart rate
bpm

Diabetes ---
If yes and on diabetic medication, please complete the Medication Module

Blood glucose concentration
mmol/L

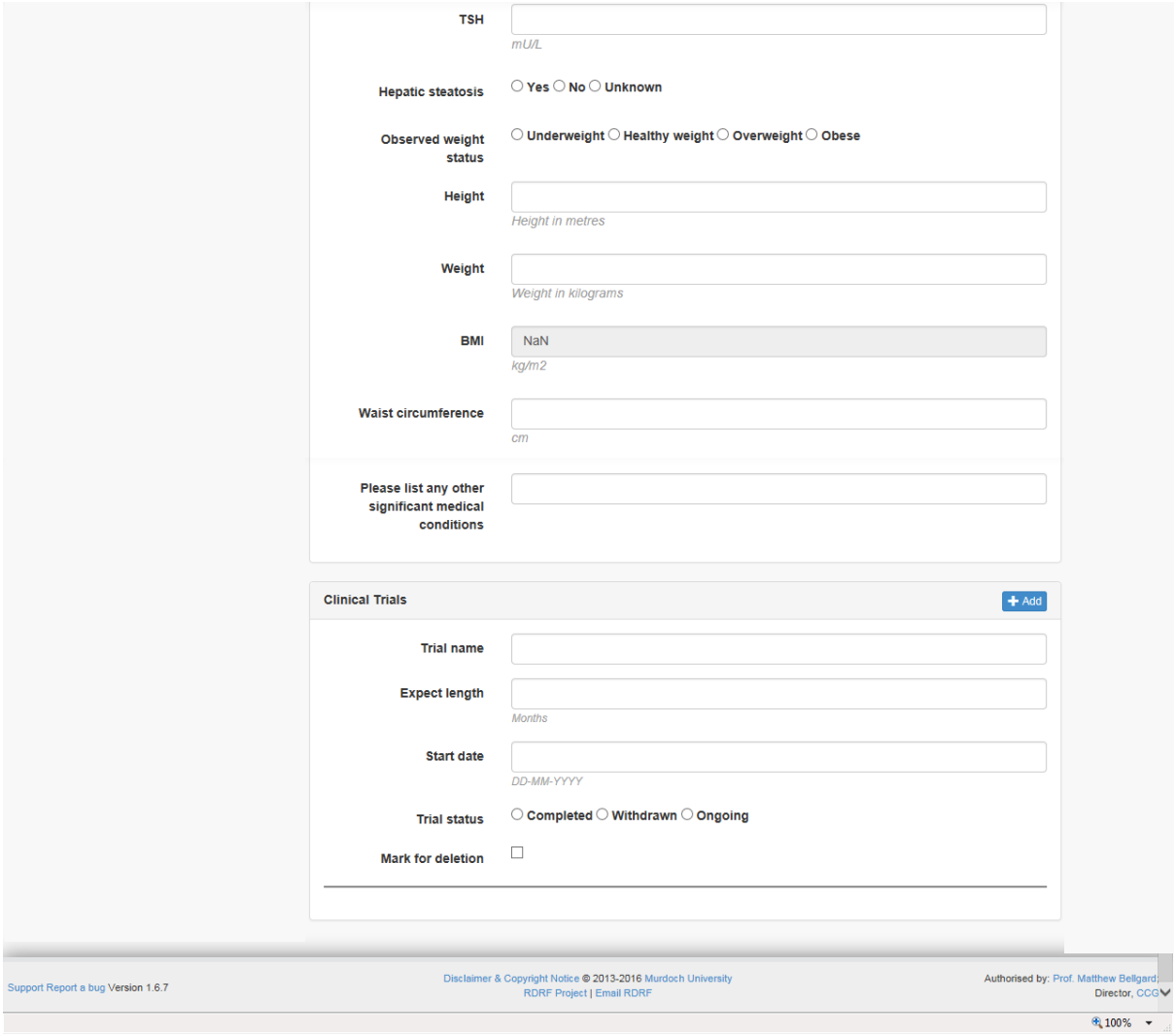
HbA1c
(%)

Chronic kidney disease ○ Yes ○ No ○ Unknown

eGFR
ml/min/1.73m2

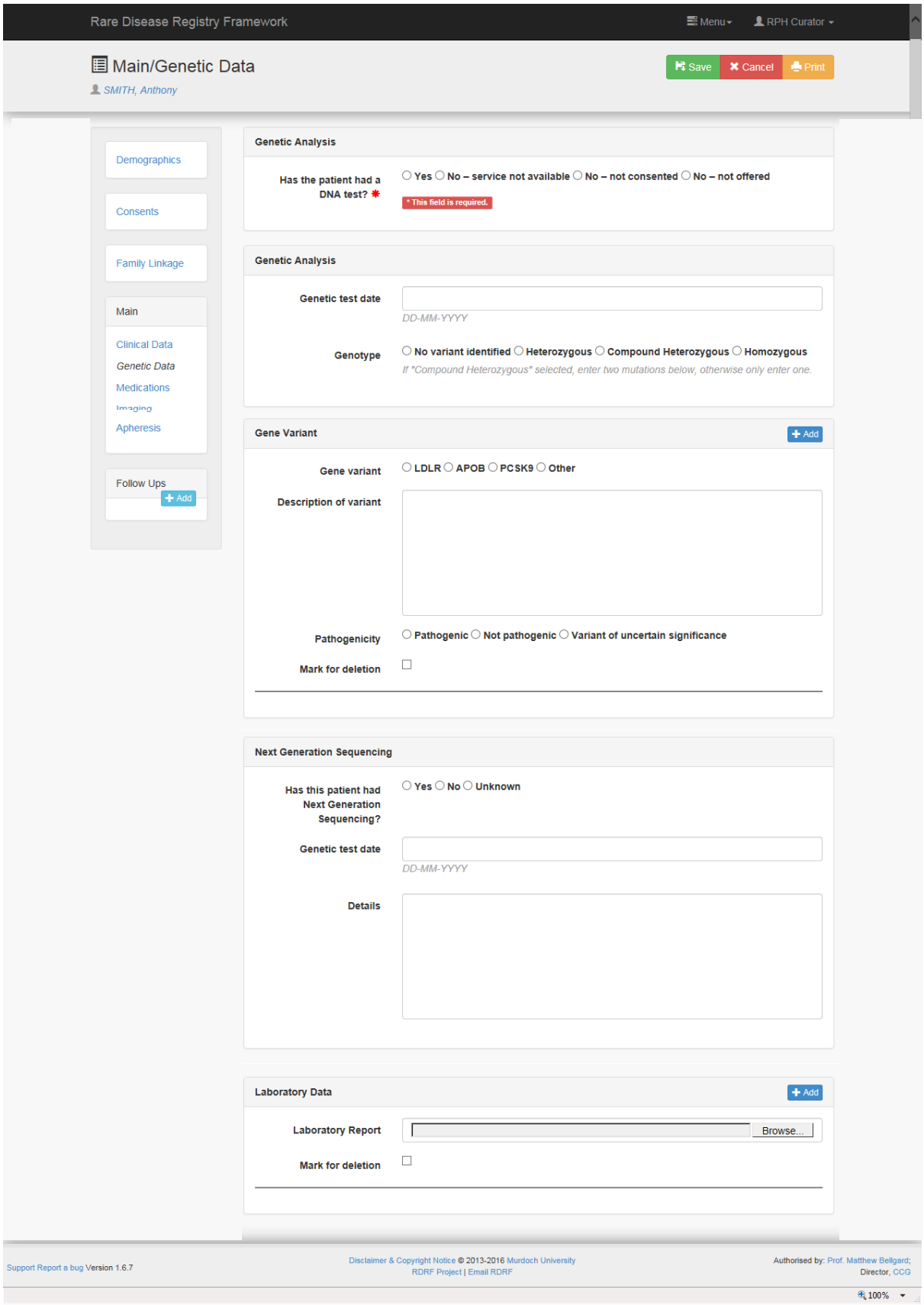
Hypothyroidism ---

RETURN TO MENU



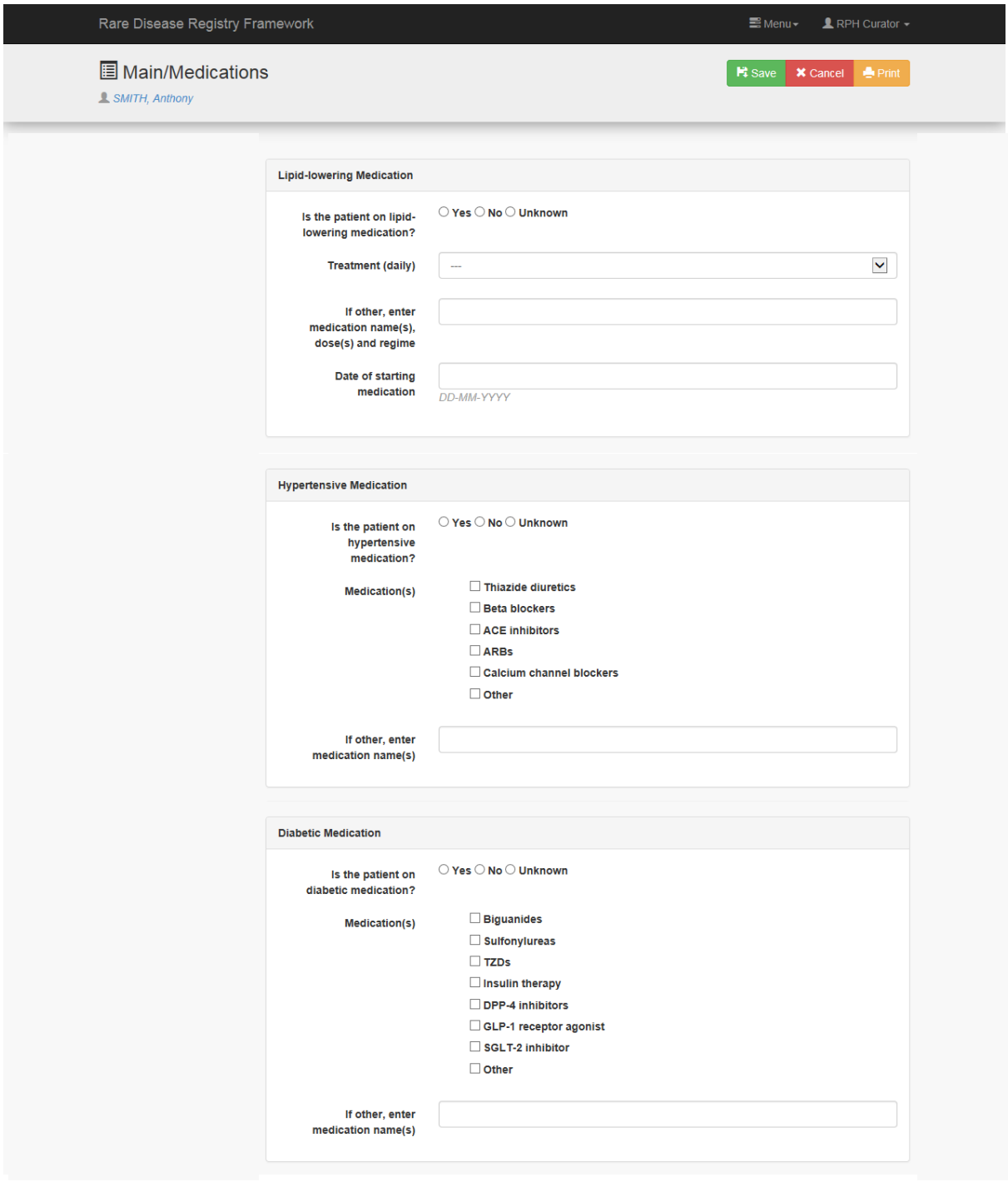
RETURN TO MENU

Screen Shot – Genetic Data (enlarge to view)



RETURN TO MENU

Screen Shot – Medications (enlarge to view)



RETURN TO MENU

Antithrombotic Medication

Is the patient on antithrombotic medication? ○ Yes ○ No ○ Unknown

Medication(s)
- ☐ Aspirin
- ☐ Other antiplatelets
- ☐ Warfarin
- ☐ Other anticoagulants

If other, enter medication name(s)

Drug Intolerance

History of drug intolerance ○ Yes ○ No ○ Unknown

Specific Lipid-lowering Drug Intolerance + Add

Date drug intolerance first observed
DD-MM-YYYY

Drug ---

Dose ---

If other, enter medication name(s), dose(s) and regime

How long was the patient on medication before intolerance was observed?
Enter a number or 'Several'

Time frame ---
Select a time frame

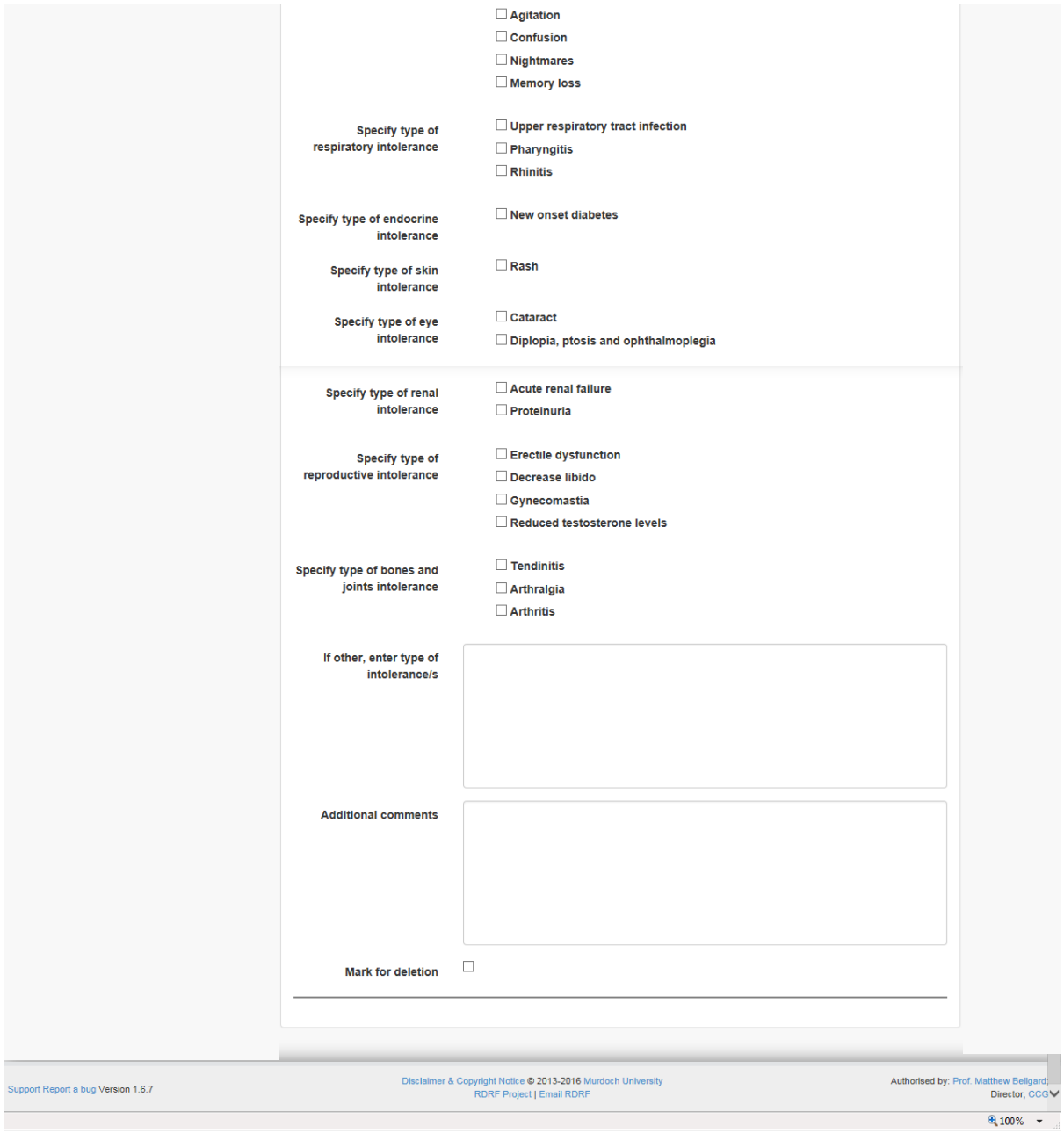
Specify type of muscle intolerance
- ☐ Fatigue
- ☐ Muscle pain
- ☐ Muscle weakness
- ☐ Muscle tenderness
- ☐ Cramps
- ☐ CK elevation
- ☐ Myalgia, tolerable
- ☐ Myalgia, intolerable
- ☐ Myopathy
- ☐ Severe myopathy
- ☐ Rhabdomyolysis
- ☐ Autoimmune-mediated
- ☐ Necrotizing myositis

Specify type of hepatic intolerance
- ☐ Hepatotoxicity
- ☐ Increased activity of liver enzymes

Specify type of gastrointestinal tract intolerance
- ☐ Constipation
- ☐ Diarrhoea
- ☐ Dyspepsia
- ☐ Flatulence
- ☐ Heartburn
- ☐ Nausea
- ☐ Vomiting

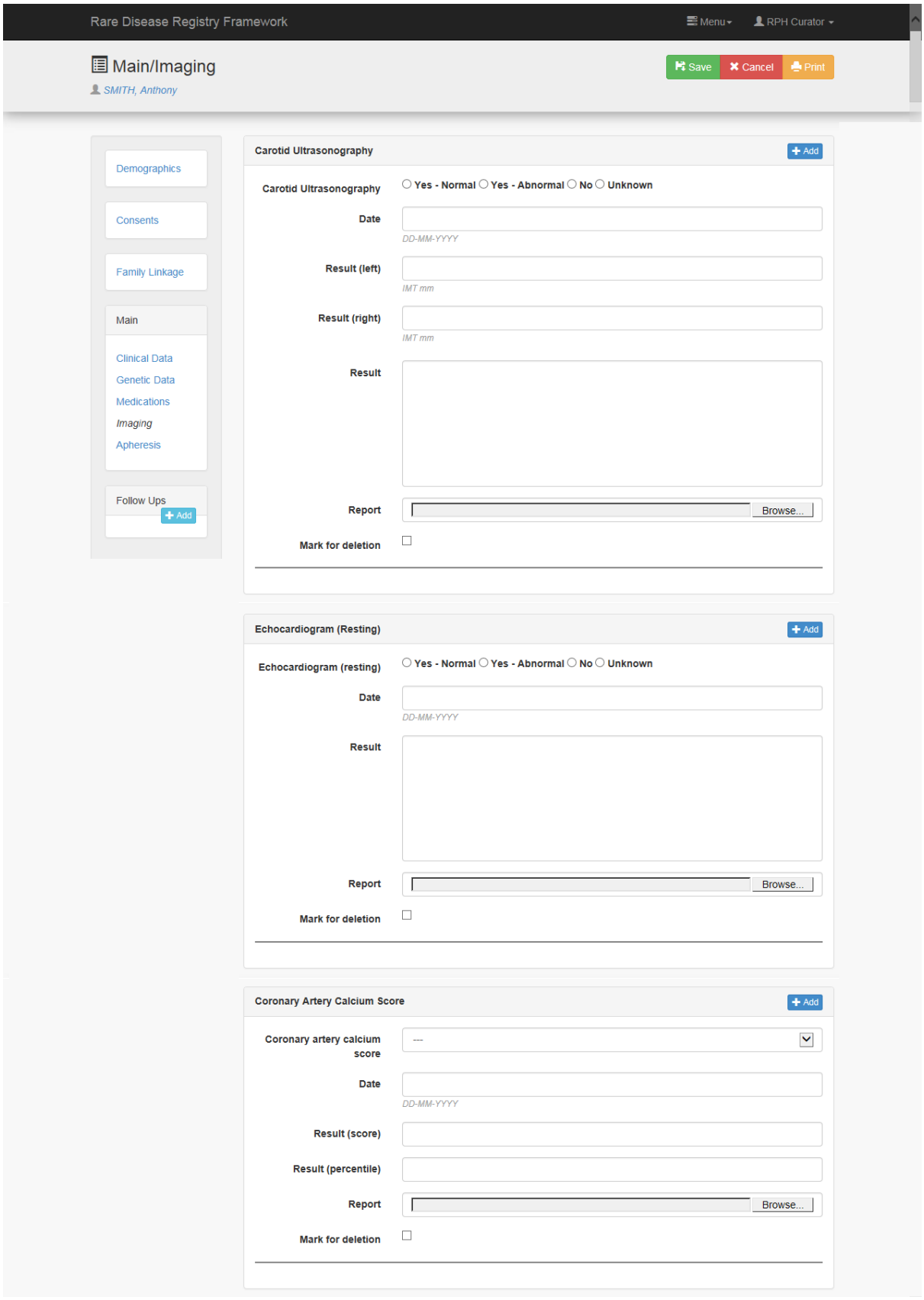
Specify type of neurologic and psychological effects intolerance
- ☐ Aggressive behaviour
- ☐ Headache
- ☐ Asthenia
- ☐ Dizziness
- ☐ Fatigue
- ☐ Severe irritability
- ☐ Insomnia
- ☐ Somnolence

RETURN TO MENU

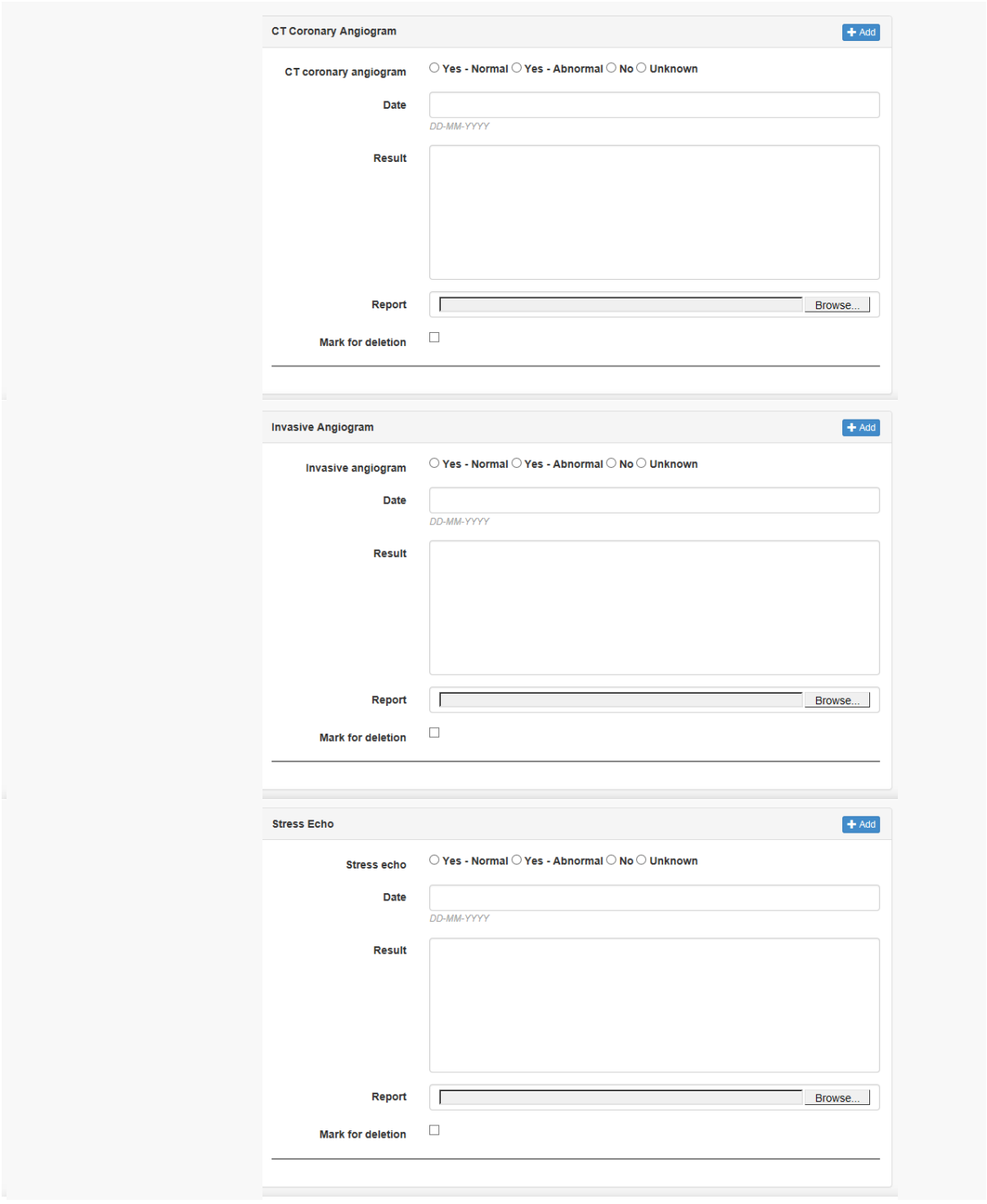


RETURN TO MENU

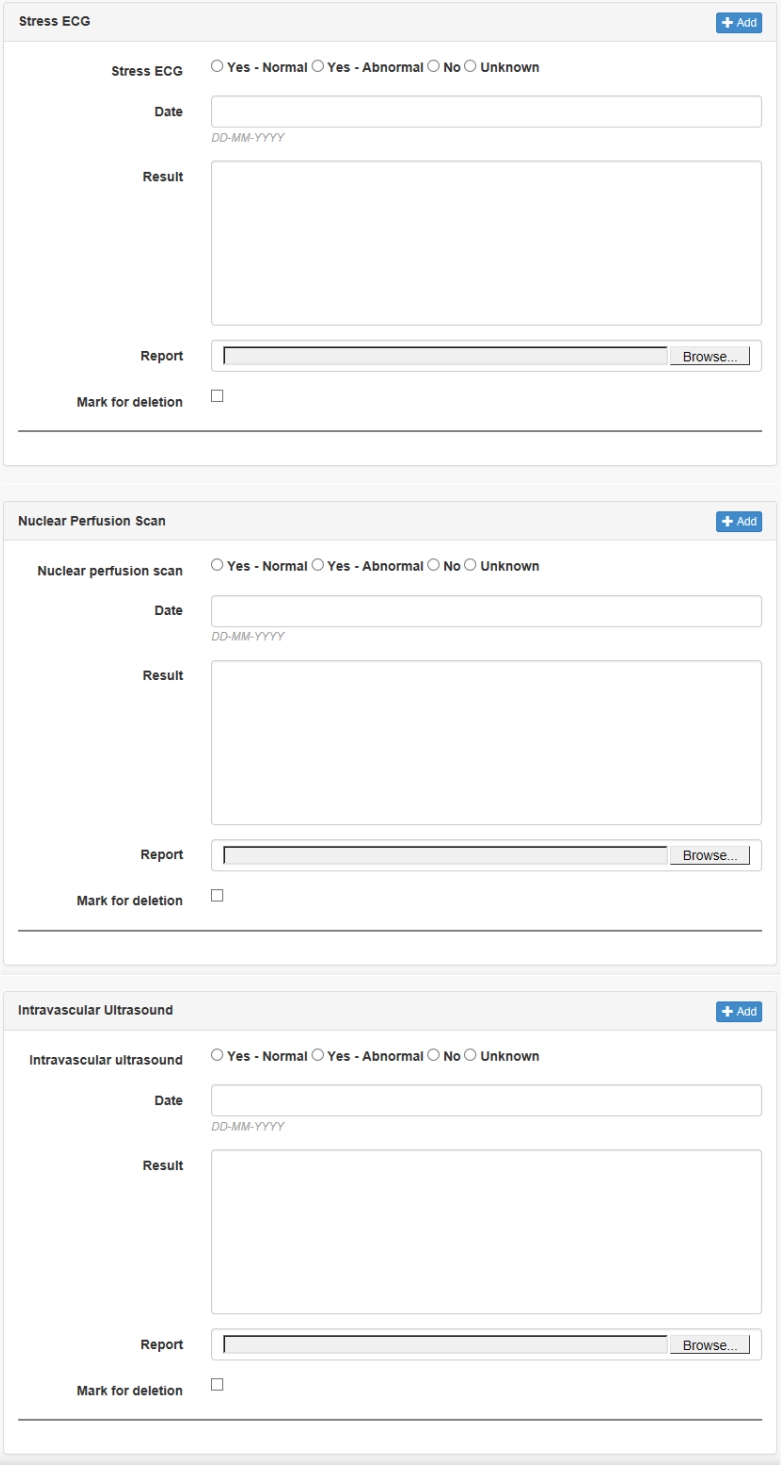
Screen Shot – Imaging (enlarge to view)



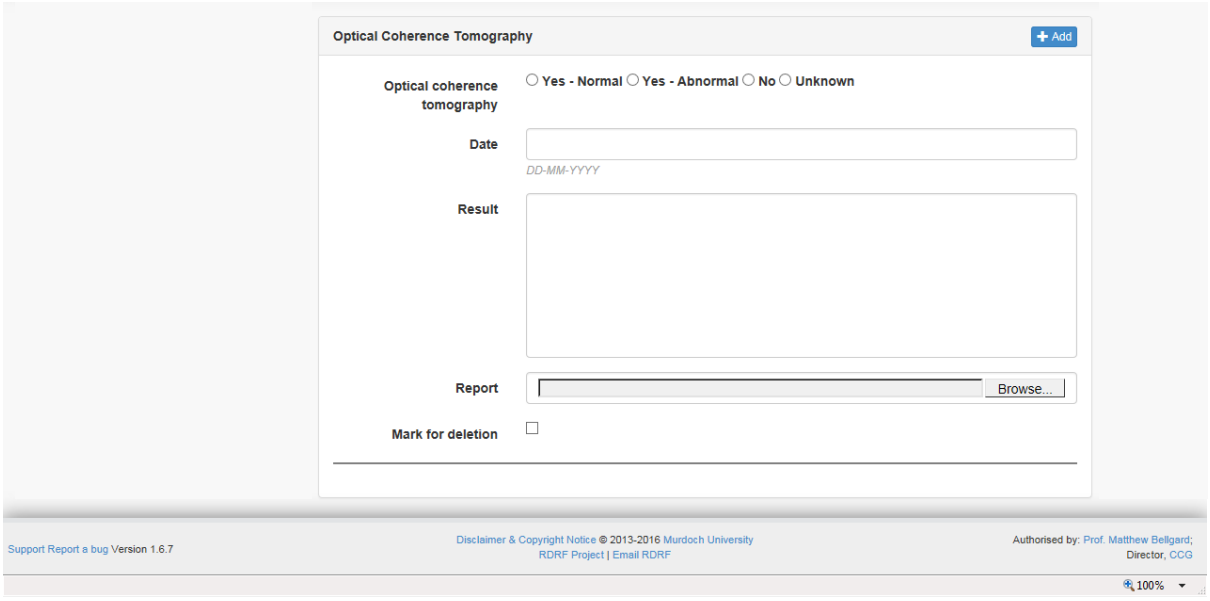
RETURN TO MENU



RETURN TO MENU

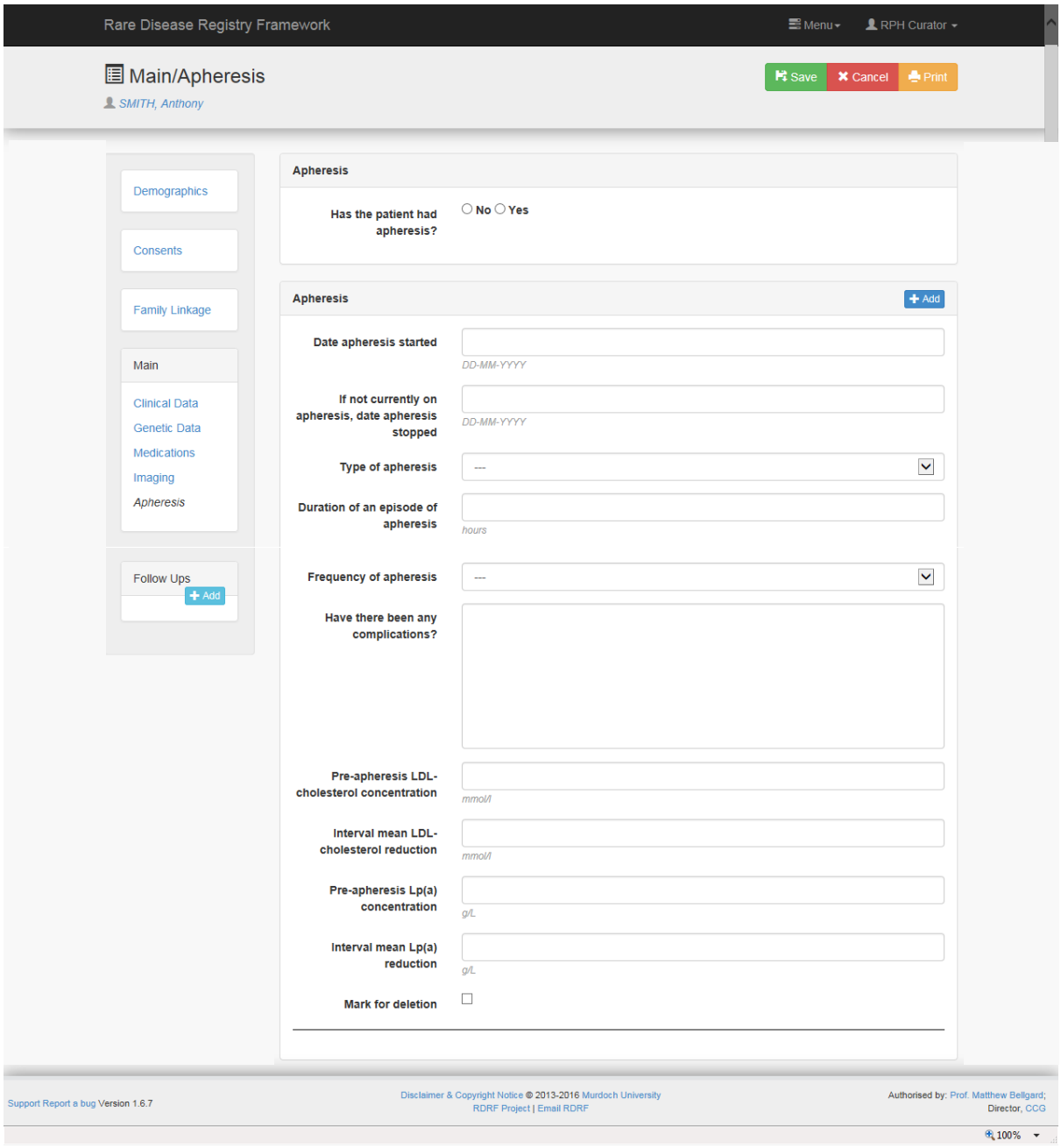


RETURN TO MENU



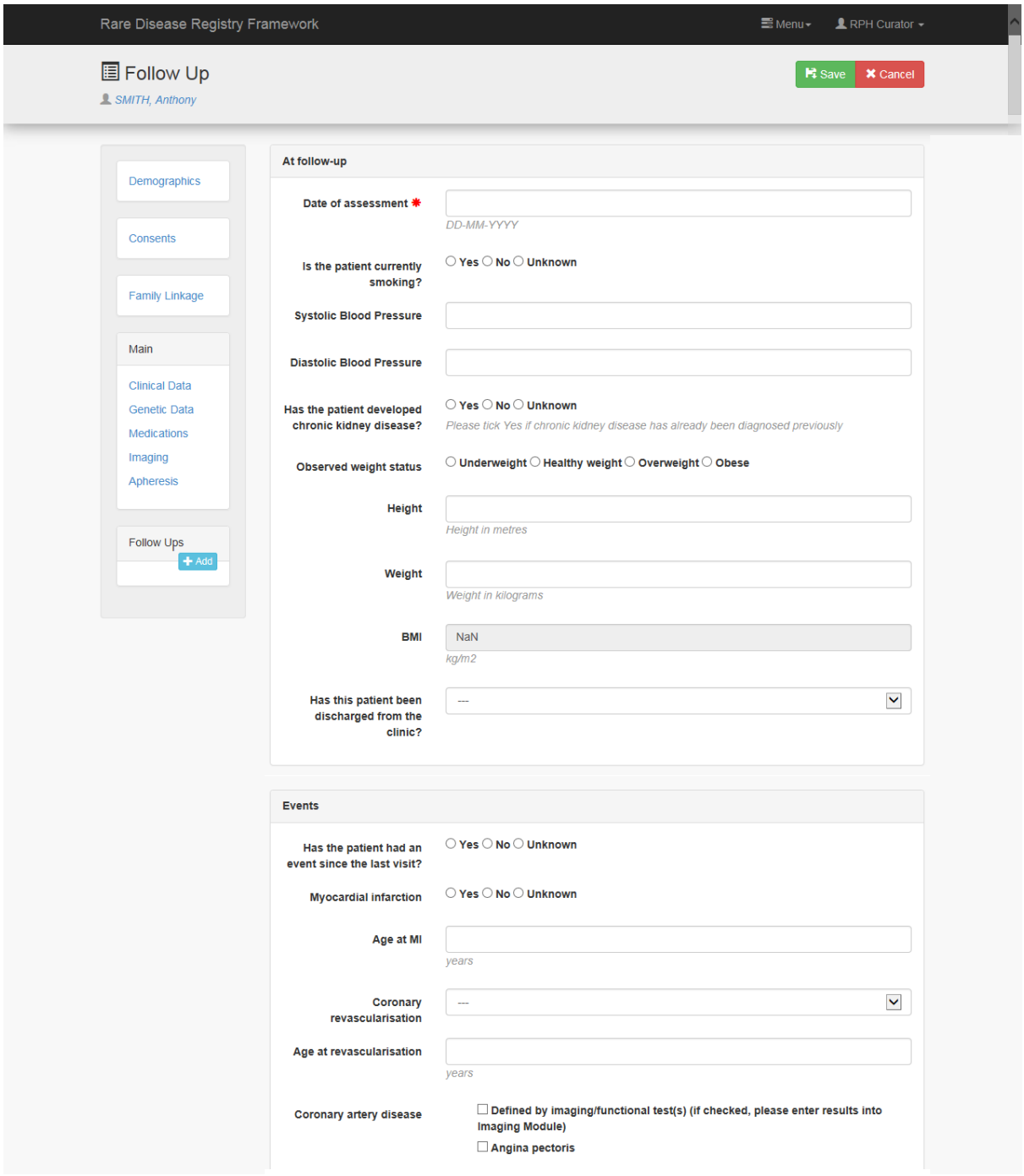
RETURN TO MENU

Screen Shot – Apheresis (enlarge to view)



RETURN TO MENU

Screen Shot – Follow Up (enlarge to view)



RETURN TO MENU

Age at angina
years

Stroke ○ Yes ○ No ○ Unknown

Age at stroke
years

TIA ○ Yes ○ No ○ Unknown

Age at TIA
years

Other vascular surgery ○ Yes ○ No ○ Unknown

Other comments
eg. multiple events or number of stents/by-passes

Hypertension

Has the patient developed hypertension? ○ Yes ○ No ○ Unknown
Please tick Yes if hypertension has already been diagnosed previously

If yes, is the patient on hypertensive medication? ○ Yes ○ No ○ Unknown

Medication(s)
- ☐ Thiazide diuretics
- ☐ Beta blockers
- ☐ ACE inhibitors
- ☐ ARBs
- ☐ Calcium channel blockers
- ☐ Other

If other, enter medication name(s)

Diabetes

Has the patient developed diabetes? ○ Yes ○ No ○ Unknown
Please tick Yes if diabetes has already been diagnosed previously

If yes, is the patient on diabetic medication? ○ Yes ○ No ○ Unknown

Medication(s)
- ☐ Biguanides
- ☐ Sulfonylureas
- ☐ TZDs
- ☐ Insulin therapy
- ☐ DPP-4 inhibitors
- ☐ GLP-1 receptor agonist
- ☐ SGLT-2 inhibitor
- ☐ Other

If other, enter medication name(s)

Antithrombotic

Has the patient been prescribed antithrombotic medication? ○ Yes ○ No ○ Unknown

Medication(s)
- ☐ Aspirin
- ☐ Other antiplatelets
- ☐ Warfarin
- ☐ Other anticoagulants

If other, enter medication name(s)

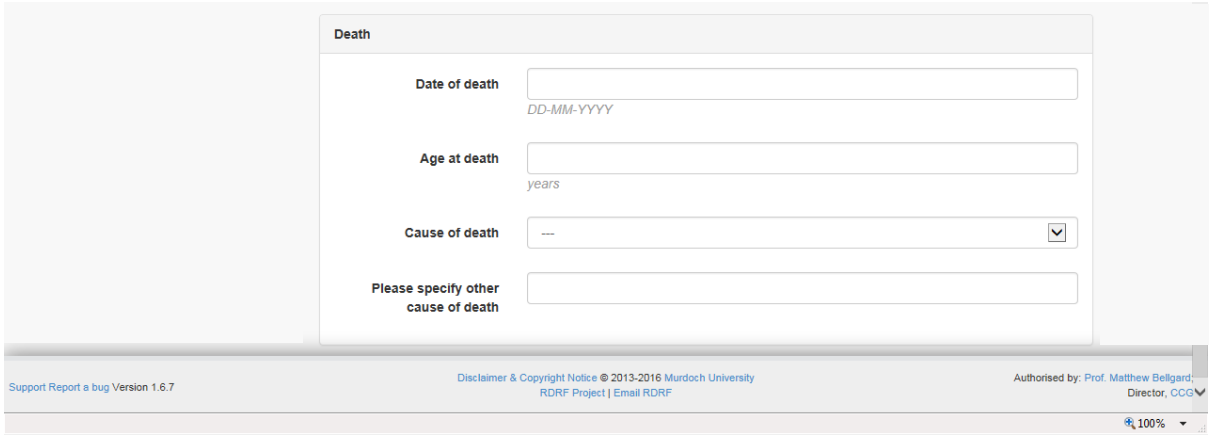
RETURN TO MENU

Biochemistry Profile

Date	DD-MM-YYYY
Total cholesterol concentration	mmol/L
LDL-cholesterol concentration	mmol/L
Triglyceride concentration	mmol/L
HDL-cholesterol concentration	mmol/L
Lp(a) concentration	g/L
Blood glucose concentration	mmol/L
HbA1c	(%)
Apolipoprotein B	g/L
Apolipoprotein A1	g/L
Aspartate transaminase (AST)	U/L
Alanine transaminase (ALT)	U/L
Creatine kinase (CK)	U/L
Albumin	g/L
Creatinine	umol/L
C-reactive protein (CRP)	mg/L
Treatment (daily)	---
If other, enter medication name(s), dose(s) and regime	
Compliance	---

If intolerant, please enter details into Medication Module - Drug Intolerance

RETURN TO MENU



RETURN TO MENU

NATIONAL fH REGISTRY

everyone deserves a better chance

Core Data Entry Form – Updated 20/7/2018

AFFIX LABEL HERE

DEMOGRAPHICS

Doctor*:	Hospital ID:
Type of Medical Professional*:	Family name*:
Home:	Given names*:
Mobile:	Address*:
Email:	PC:
Ethnic origin:	Sex*: DOB*:

☐ INDEX ☐ RELATIVE – Name of index/relationship:

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

CONSENT

☐ Adult ☐ Child ☐ Clinical trials ☐ Information ☐ FCHL ☐ Hyper-Lp(a) ☐ Upload consent file

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

CLINICAL DATA

Date of consent*: Date of assessment*:

CLINICAL DATA – FH Score		Score	Patient Score
1.0	**Family History**		
1.1	First degree relatives with known premature coronary and vascular disease (Men < 55 years, Females < 60 years) **Father / Mother / Other:**_____	**1**	
1.2	**OR** First degree relatives with known LDL-cholesterol (LDL-C) above the 95th percentile (for age and sex) **Father / Mother / Other:**_____		
1.3	First degree relatives with ☐ tendinous xanthomata and/or ☐ arcus cornealis	**2**	
1.4	**OR** Have children aged less than 18 years with LDL-C above the 95th percentile (for age and sex)		
2.0	**Clinical History**		
2.1	Patient with premature coronary artery disease (Men < 55 years, Females < 60 years) **MI: age at first**_____ **CABG: age at first**_____ **PCI: age at first**_____	**2**	
2.2	Patient with premature cerebral or peripheral vascular disease (Men < 55 years, Females < 60 years) **Age at first**_____	**1**	
3.0	**Physical examination**		
3.1	Tendinous xanthomata **Right / Left / Bilateral**	**6**	
3.2	Arcus cornealis prior to age 45 years **Right / Left / Bilateral**	**4**	
4.0	**Low Density Lipoprotein Cholesterol, LDL-C (mmol/L)**		
4.1	LDL-C ≥ 8.5	**8**	
4.2	LDL-C 6.5-8.4	**5**	
4.3	LDL-C 5.0-6.4	**3**	
4.4	LDL-C 4.0-4.9	**1**	

☐ UNTREATED LDL:_____mmol/L
OR
☐ TREATED LDL:_____mmol/L
Treatment:
LDL adjusted for treatment_____mmol/L

FH Diagnostic Categories: Definite >8, Probable 6-8, Possible 3-5, Unlikely 0-2

FH Score:

Plasma LDL-cholesterol for FH Score

HIGHEST UNTREATED LDL-C*
OR TREATED LDL-C and Treatment
(Closest to the 'Date of consent')

CLINICAL DATA – Biochemistry Profile *Enter one profile closest to the 'Date of consent'. This profile can be treated or untreated.*

Date*		
Total cholesterol*:	LDL-cholesterol*:	Treatment (daily)*: *(At time of lipid profile)*
Triglyceride*:	HDL-cholesterol*:	
Compliance*	☐ Full ☐ Irregular ☐ Non-compliant ☐ Intolerant ☐ N/A ☐ Unknown	

CLINICAL DATA – Other CVD Risk Factors

Smoking*	☐ Yes ☐ Never ☐ Ex-smoker ☐ Unknown	
Alcohol*	☐ Yes – daily ☐ Yes – once/twice a week ☐ Yes – occasional ☐ Never ☐ Former ☐ Unknown	
Hypertension*	☐ Yes ☐ No ☐ Unknown	Systolic: Diastolic:
Diabetes*	☐ Yes ☐ No ☐ Unknown	Glucose: HbA1c (%):
Chronic kidney disease*	☐ Yes ☐ No ☐ Unknown	eGFR:
Hypothyroidism*	☐ Yes – treated ☐ Yes – untreated ☐ No ☐ Unknown	TSH:
Observed weight status*	☐ Underweight ☐ Healthy weight ☐ Overweight ☐ Obese ☐ Unknown	
	Height (m):	Weight (kg):
Lp(a)*		
Other significant medical conditions:		

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

GENETIC DATA

DNA test*	☐ Yes ☐ No – service not available ☐ No – not consented ☐ No – not offered *If yes, complete sections below.*
Date	
Genotype	☐ None identified ☐ Hetero. ☐ Compound Hetero. ☐ Homozygous
Gene Variant	☐ LDLR ☐ APOB ☐ PCSK9 ☐ Other
Description	
Pathogenicity	☐ Pathogenic ☐ Non-pathogenic ☐ Uncertain ☐ Upload report

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

MEDICATIONS

Lipid-lowering medication* *(At time of entry into the registry)*	☐ Yes ☐ No ☐ Unknown *If yes, complete below:* Treatment (daily)*: Date of first starting any lipid-lowering medication*:
Hypertensive medication*	☐ Yes ☐ No ☐ Unknown *If yes, complete below:* ☐ Thiazide diuretics ☐ Beta blockers ☐ ACE inhibitors ☐ ARBs ☐ Calcium channel blockers ☐ Other
Diabetic medication*	☐ Yes ☐ No ☐ Unknown *If yes, complete below:* ☐ Biguanides ☐ Sulfonylureas ☐ TZDs ☐ Insulin therapy ☐ DPP-4 inhibitors ☐ GLP-1 receptor agonist ☐ SGLT-2 inhibitor ☐ Other
Antithrombotic medication*	☐ Yes ☐ No ☐ Unknown *If yes, complete below:* ☐ Aspirin ☐ Other antiplatelets ☐ Warfarin ☐ Other anticoagulants
History of drug intolerance*	☐ Yes ☐ No ☐ Unknown *If yes, complete more details. Enter as much detail as is available.*

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

IMAGING *Not all imaging is listed here. Please complete all imaging in the registry.*

Carotid ultrasonography*	☐ Yes - Normal ☐ Yes - Abnormal ☐ No ☐ Unknown *If yes, complete more details.*
Echocardiogram (resting)*	☐ Yes - Normal ☐ Yes - Abnormal ☐ No ☐ Unknown *If yes, complete more details.*
Coronary artery calcium score*	☐ Yes-0 ☐ Yes-1-99 ☐ Yes-100-300 ☐ Yes->300 ☐ No ☐ Unknown *If yes, complete more details.*
CT coronary angiogram*	☐ Yes - Normal ☐ Yes - Abnormal ☐ No ☐ Unknown *If yes, complete more details.*
Invasive angiogram*	☐ Yes - Normal ☐ Yes - Abnormal ☐ No ☐ Unknown *If yes, complete more details.*
Other:	☐ Yes - Normal ☐ Yes - Abnormal ☐ No ☐ Unknown *If yes, complete more details.*

Click the green 'Save' button, then use the blue arrow on the right to move to the next form >

APHERESIS*	☐ Yes ☐ No ☐ Unknown *If yes, complete more details.*

NATIONAL fH REGISTRY

everyone deserves a better chance

AFFIX LABEL HERE

DEMOGRAPHICS	Hospital ID:
Dr:	Family name:
Date*:	Given names:
Home:	Address:
Mobile:	PC: Sex: DOB:
Email:	
☐ **INDEX** ☐ **RELATIVE** Index/relationship:	

Currently smoking: ☐ Yes ☐ No ☐ Unknown		
Hypertension: Systolic Blood Pressure:	Diastolic Blood Pressure:	
Chronic kidney disease: ☐ Yes ☐ No ☐ Unknown	Tick yes if chronic kidney disease has been diagnosed previously.	
Observed weight status: ☐ Underweight ☐ Healthy weight ☐ Overweight ☐ Obese		
Height (m):	Weight (kg):	BMI:
Patient discharged: ☐ Yes – to GP ☐ Yes – to specialist ☐ No – remains in clinic ☐ No – but lost to follow up ☐ Unknown		

EVENTS	
Event since last visit: ☐ Yes ☐ No ☐ Unknown	
Myocardial infarction: ☐ Yes ☐ No ☐ Unknown	Age at MI (years):
Coronary revascularisation: ☐ PCI ☐ CABG ☐ Both ☐ No ☐ Unknown	Age at revascularisation (years):
Coronary artery disease: ☐ Defined by imaging/functional test(s)	If checked enter results into Imaging Module.
☐ Angina pectoris	Age at angina (years):
Stroke: ☐ Yes ☐ No ☐ Unknown	Age at stroke (years):
TIA: ☐ Yes ☐ No ☐ Unknown	Age at TIA (years):
Other vascular surgery: ☐ Yes ☐ No ☐ Unknown	
Other comments e.g. multiple events or number of stents/by-passes:	

HYPERTENSION	
Hypertension: ☐ Yes ☐ No ☐ Unknown	Tick yes if hypertension has been diagnosed previously.
Hypertensive medication: ☐ Yes ☐ No ☐ Unknown	
Medications: ☐ Thiazide diuretics ☐ Beta blockers ☐ ACE inhibitors ☐ ARBs ☐ Calcium channel blockers ☐ Other	
If other, enter medication name(s):	

DIABETES			
Diabetes: ☐ Yes ☐ No ☐ Unknown	Tick yes if diabetes has been diagnosed previously.		
Diabetic medication: ☐ Yes ☐ No ☐ Unknown			
Medications: ☐ Biguanides	☐ Sulfonylureas	☐ TZDs	☐ Insulin therapy
☐ DPP-4 inhibitors	☐ GLP-1 receptor agonist	☐ SGLT-2 inhibitor	☐ Other
If other, enter medication name(s):			

ANTITHROMBIC

Antithrombic medication: ☐ Yes ☐ No ☐ Unknown

Medications: ☐ Aspirin ☐ Other antiplatelets ☐ Warfarin ☐ Other anticoagulants

If other, enter medication name(s):

BIOCHEMISTRY PROFILE

Date	
TC	
LDL	
TG	
HDL	
Lp(a)	
Glucose	
HbA1c	
ApoB	
ApoA1	
AST	
ALT	
CK	
Alb	
Creat	
CRP	

CURRENT MEDICATION (Lipid lowering)

Name	Dose	Frequency	Commenced

Compliance: ☐ Full ☐ Irregular ☐ Non-compliant ☐ Intolerant ☐ N/A

Details if intolerant - Enter details into Medication Module – Drug intolerance:

DEATH

Date of death: | Age at death (years):

Cause of death: ☐ CAD ☐ Cerebrovascular disease - ischemic ☐ Cerebrovascular disease - haemorragic ☐ Cardiac sudden death ☐ Heart failure ☐ Acute aortic syndrome ☐ Cancer ☐ Other

Specify other cause of death:

ADDITIONAL NOTES