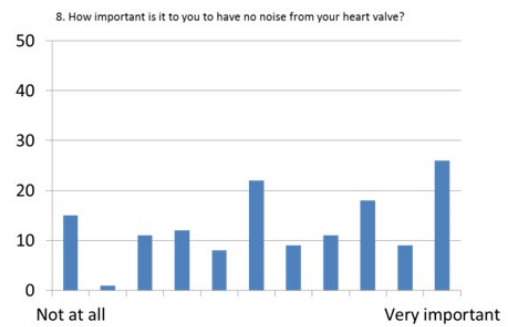
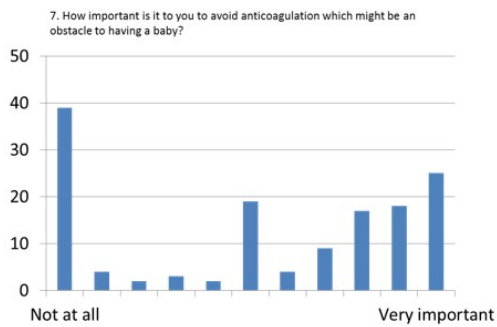
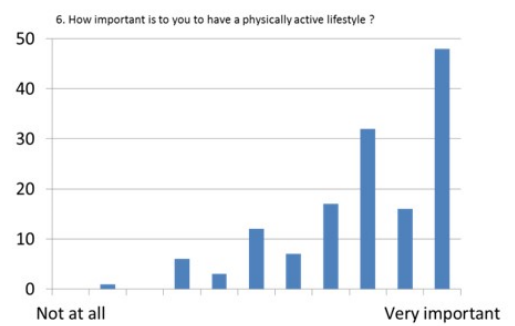
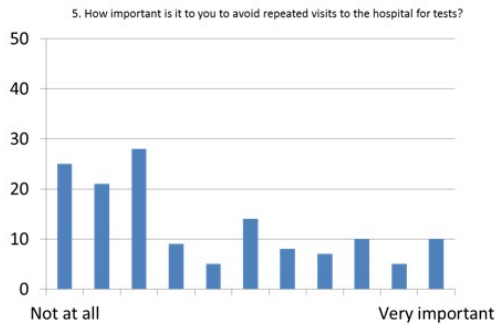
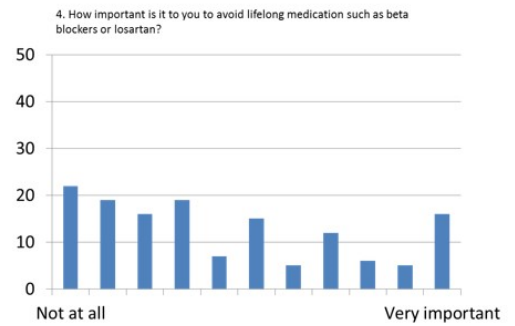
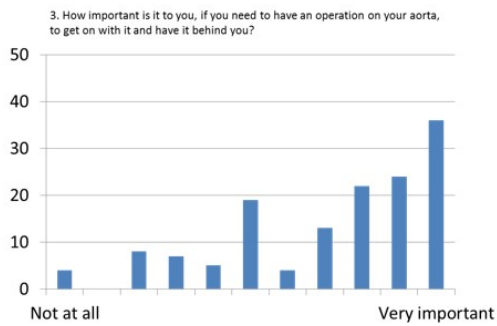
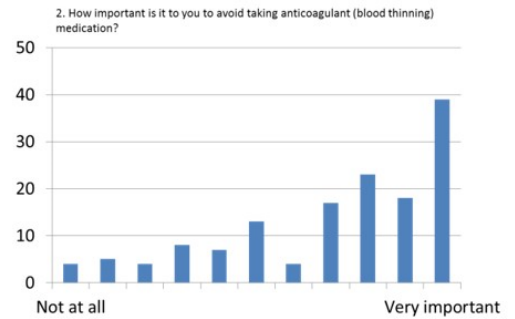
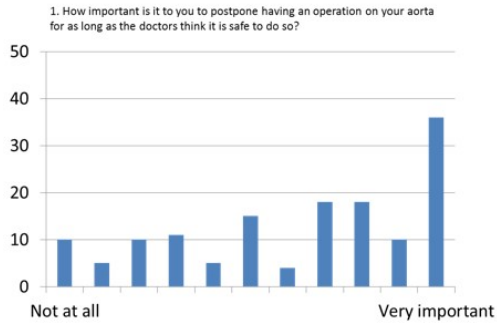


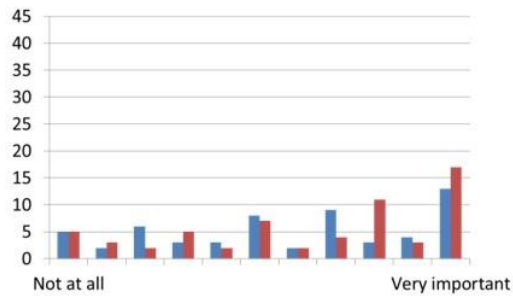
# Supplement

## 1. Responses for all 142 respondents

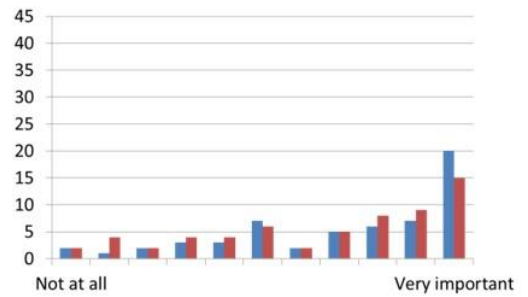


## 2. Males (N=58 blue) versus females (N=61 red) of 119 people with Marfan syndrome (including two doctors)

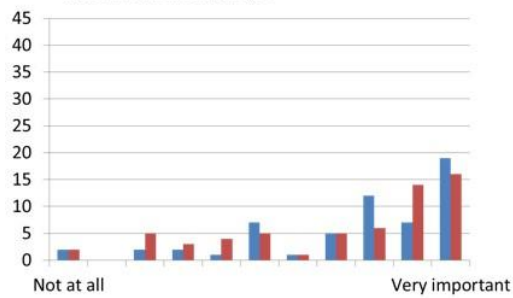
1. How important is it to you to postpone having an operation on your aorta for as long as the doctors think it is safe to do so?



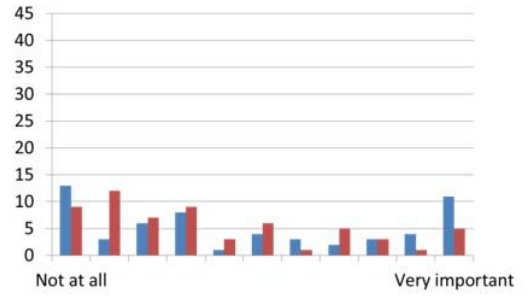
2. How important is it to you to avoid taking anticoagulant (blood thinning) medication?



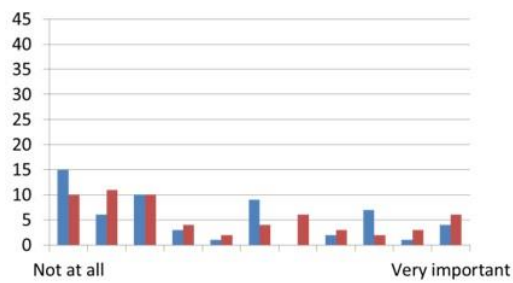
3. How important is it to you, if you need to have an operation on your aorta, to get on with it and have it behind you?



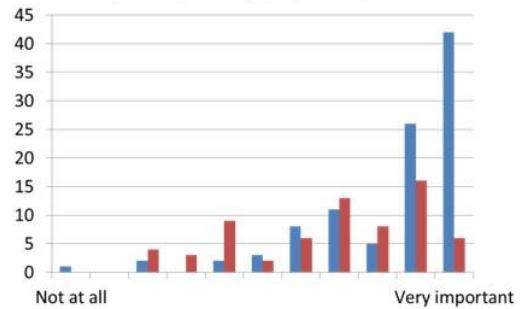
4. How important is it to you to avoid lifelong medication such as beta blockers or losartan?



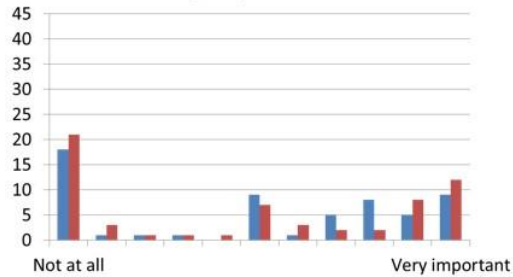
5. How important is it to you to avoid repeated visits to the hospital for tests?



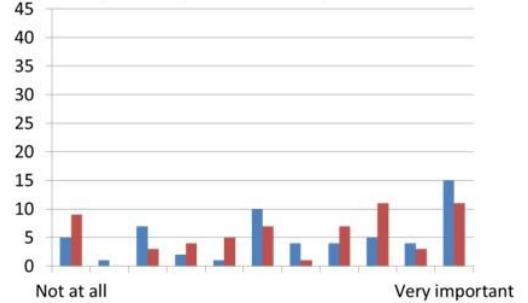
6. How important is it to you to have a physically active lifestyle?



7. How important is it to you to avoid anticoagulation which might be an obstacle to having a baby?

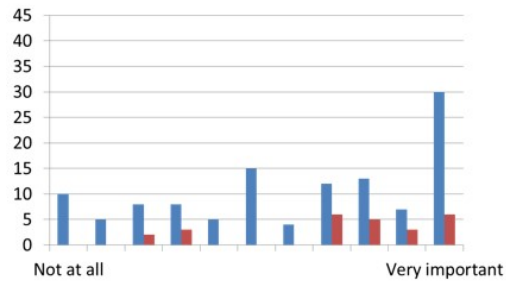


8. How important is it to you to have no noise from your heart valve?

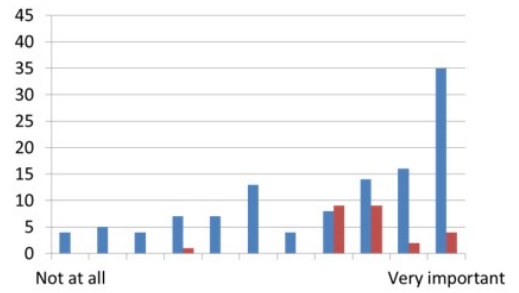


### 3. People affected by Marfan syndrome (N=117, blue) compared with doctors (N=25 red) including the 2 who also had Marfan syndrome).

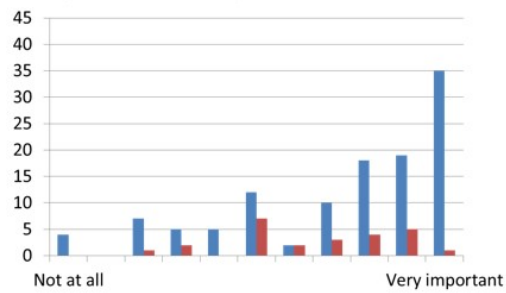
1. How important is it to you to postpone having an operation on your aorta for as long as the doctors think it is safe to do so?



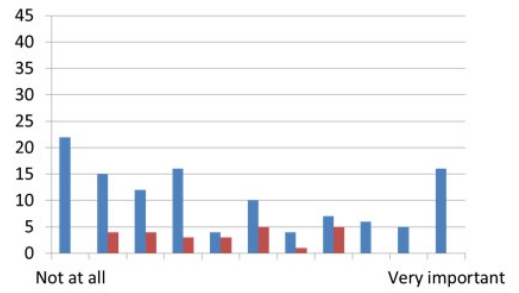
2. How important is it to you to avoid taking anticoagulant (blood thinning) medication?



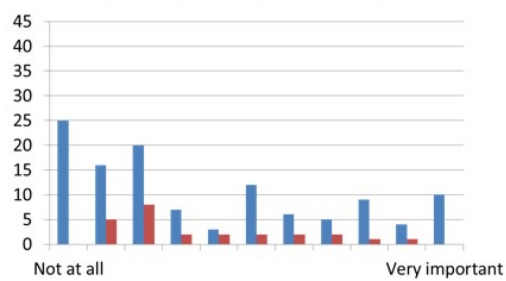
3. How important is it to you, if you need to have an operation on your aorta, to get on with it and have it behind you?



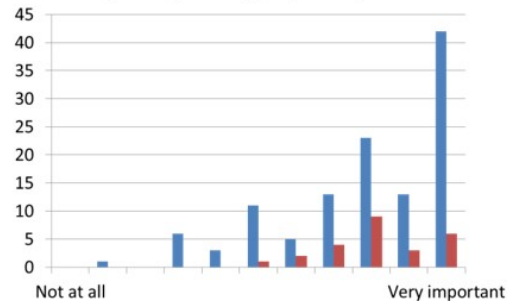
4. How important is it to you to avoid lifelong medication such as beta blockers or losartan?



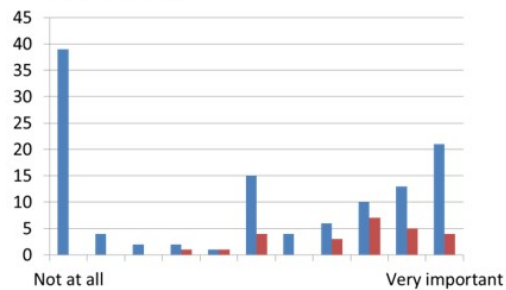
5. How important is it to you to avoid repeated visits to the hospital for tests?



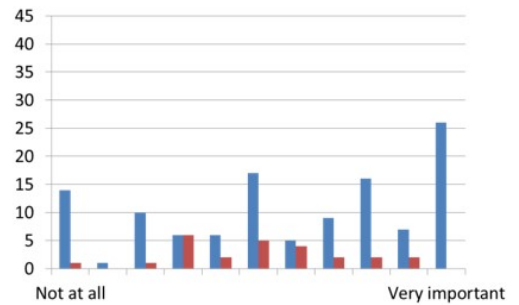
6. How important is it to you to have a physically active lifestyle?



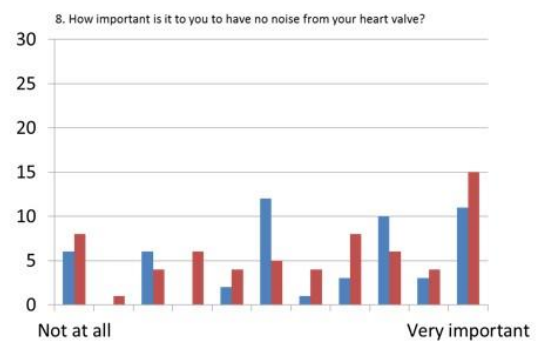
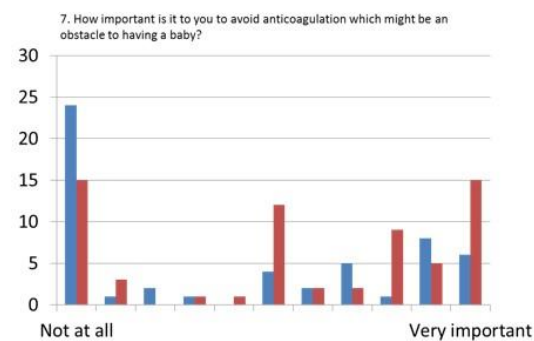
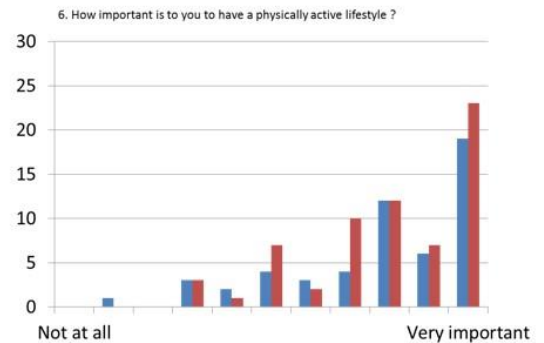
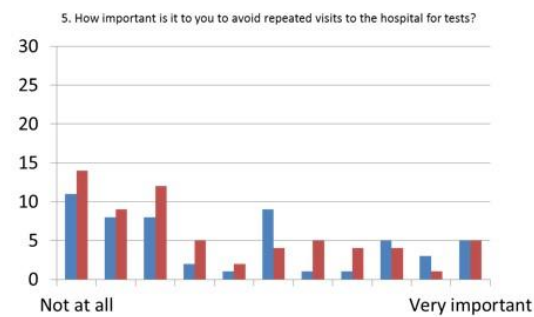
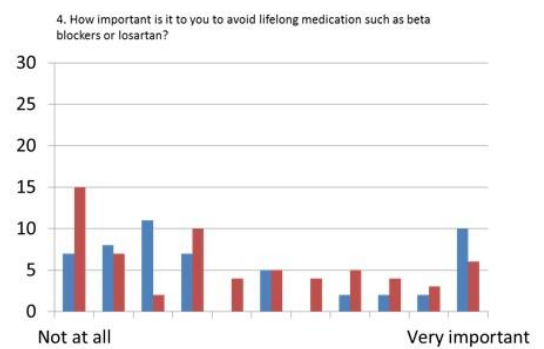
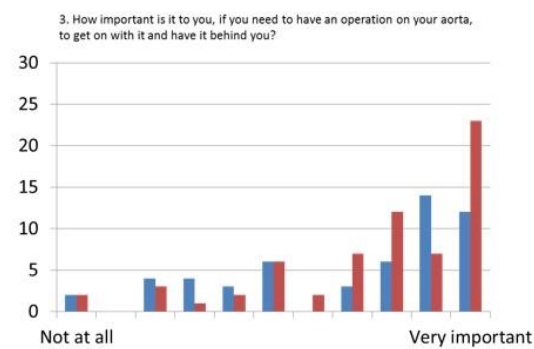
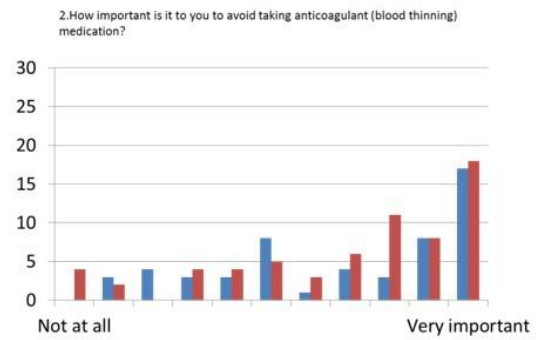
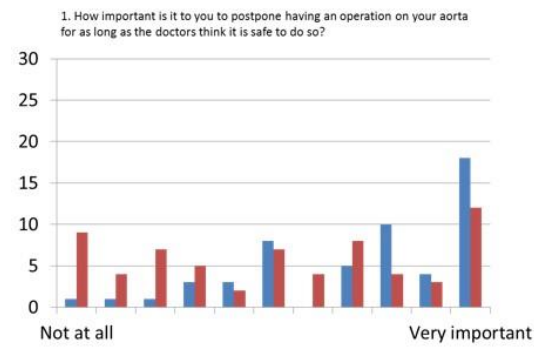
7. How important is it to you to avoid anticoagulation which might be an obstacle to having a baby?



8. How important is it to you to have no noise from your heart valve?



## 4. People who had already had root surgery (N=65 red) were compared with the 54 who were potential candidates (N=54 blue).



## 5. Findings from the analysis of the free text boxes

The free text responses in the questionnaire related closely to the overall choices respondents made when rating the importance of the items they were asked about. Responses were often based on personal experiences, or those of relatives, and hence varied. Where direct personal experience was absent, many respondents nevertheless identified with the decision making dilemma presented in the question and took a viewpoint. An example of this was the commentary to question seven: “How important is it to you to avoid anticoagulation which might be an obstacle to having a baby?” Men, and also women beyond child bearing age, empathised with the dilemma in making a decision about starting a family when faced with heightened risks to the pregnancy through anticoagulation medication. One female respondent who had her children before she was diagnosed with Marfan placed a value of ten on avoiding anticoagulation as her response and elaborated as follows: “I have always been grateful that I had already had my child before diagnosis took place - thus no decision was necessary...” (Respondent 12, age 67).

Doctors tended to consider the issues from a broad psycho-social perspective, highlighting the impact of treatment choices and decisions on life style, life course, and the patient’s ability to manage the condition. For example, some of the doctors pointed out that anticoagulation medication not only can lead to complications, but also requires particular life style choices. Hence, decisions need to take the personal circumstances of the patient into account: “Having a young patient I believe it is important to offer options that will not include use of Warfarin” (Respondent 110, doctor); “May seem less of a problem as one ages (engaging in less physical activity)” (Respondent 8, doctor).

### Recurring themes from all respondents

Recurring themes informing scores and answers across the questionnaire were identified as

- Maintaining as normal a life as possible;
- Reserving a good quality of life in living with Marfan syndrome; and
- Retaining an active and participatory lifestyle.

Preserving a normal life for as long as possible was particularly important in the decision to delay aortic root surgery. Commentary in favour of delay included considerations of risks of the procedure and outcome; recovery from the operation; current (good) health; and the current life course status, such as having a young family; and existing work/study commitments. Respondents in favour of having the procedure sooner rather than later commented on the disruption and levels of anxiety experienced in waiting for the procedure and the uncertainty delay posed to moving forward with one's life.

While patients were prepared to live with some restrictions in managing their condition, their viewpoints were informed by the wish to maintain a good quality of life. While taking life-time medication may be an inconvenience, it was considered acceptable to many respondents because they perceive it as a condition for maintaining their current health status for longer. Conversely, for some respondents, the reluctance to take medication influenced the decision to have an operation. Where the quality of life was seen to be better served by having an operation, then this was also highlighted as a reason to proceed with it.

The wish to preserve an active life style was a common theme throughout the commentary to all the questions and in many instances informed decision-making about surgery. Being active not only included being physically active, but was also highlighted as being able to participate in everyday life. In particular, anticoagulation medication was viewed by many respondents as impacting negatively on being able to retain an active life style.

The preferences and sometimes strong views offered in the study were tempered by a sense of pragmatism in dealing with some of the treatments that come with the management of Marfan syndrome. People on long-term medication commented on the impact that these may have on everyday life, including the impact of having to manage side effects of medication. In the main, respondents were accepting it as part of living with Marfan syndrome. "You learn to cope with it" (Respondent 54, male, age 68); "I do not worry about it" (Respondent 48, female, age 45).

### **The perspective of doctors**

The medical practitioners in the study commented on the various questions from the perspective of 'risks' involved, and highlighted the dilemma the questions posed for the various decisions in managing Marfan. Risks included "risks of surgery against risk of rupture" (Respondent 08, doctor); risks of complications through medication; risks of affecting physical functioning (for example: the risk of impotence); plus the impact on life course decisions such as pregnancy.

When considering whether or not to postpone surgery, most of the doctors took the view that surgery should only be conducted when it became medically necessary because of the risks associated with the surgery. The exception to this was the view of one doctor, who made the case for earlier surgical intervention: "Fix the problem may signify return to normal life" (Respondent 1 doctor).

In relation to the three dominant themes identified for respondents with Marfan, maintaining an active life style was the one highlighted by doctors as a decisive factor. Doctors cited examples of being active in everyday life, whether that was walking, or keeping physical active through other activities, as a yardstick for deciding whether a surgical procedure would be most appropriate. It may be that the value placed on remaining active, especially physically active, is most accessible to the outside view.