

**SUPPLEMENTAL FILE 6.** Coding tree and full version results of in-depth interviews.

In the third phase of the content analysis procedure of all 11 interviews, SD and EG analyzed the codes together to construct the narrative syntheses (i.e., selective coding) and the final coding tree.

**Coding tree**

- Psychological distress
  - (Fairly) No psychological distress
  - Moderate level of psychological distress
  - Prominent level of psychological distress
- Communication
  - Communication within healthcare team
  - Communication within familial context
- Sense of control
  - Sense of control through disease-related knowledge
  - Sense of control in unpredictable and uncertain prognosis
- Coping styles
  - Coping style in relation to the self
  - Coping style in relation to partner, family and environment
- Expressions of depressive feelings because of the condition
- Physical complaints
- Sense of loss
  - Sense of loss in daily life, spare time and social life
  - Sense of loss in personality and professional choice
  - Sense of loss in future and thoughts about family planning.

Figure 4 is based on the results of this coding tree. In this tree the pronunciations of the interviewees were divided under the different subjects. These pronunciations were analysed and summarized in the result section below.

**Result section**Psychological distress rather than feelings of fear

Fear did not appear as the main topic in our analyses as was hypothesized at the start of this study. All participants did, however, express some level of emotional (di)stress related to their condition, although a large variability between interviewees was observed. While some patients expressed feelings of being overwhelmed by the condition and experienced daily, persistent and paralyzing emotional distress, others only encountered distress when an outpatient check-up was approaching. Hence, the concept of psychological/emotional distress is deemed more in line with patients' expression in contrast to the concept of 'fear'. Furthermore, three different groups could be described in our sample based on their

respective level of experienced psychological distress: (i) (fairly) no psychological distress; (ii) moderate level of psychological distress; and (iii) prominent level of psychological distress.

The majority of patients (n=5/11) in our sample, expressed (fairly) no psychological distress. The explanation of this absence of fear appeared to be found in relation to regularly check-ups performed at the outpatient clinic, the confirmation of a stable aorta diameter and a specific, down-to-earth and rational personality. The only time these patients reported some distress was a short period before an outpatient check-up. Within this group, patients did not report having fear of being alone or getting complications after an operation in the future. Furthermore, they did not spontaneously express any concerns about the hereditary nature of their condition in relation to their children.

Three participants experienced variable levels of distress at different points in time (i.e., moderate level of psychological distress). Distress was not experienced on a daily basis, nor was it predominant in patients' daily. One patient said: *"It's fine that I know my diagnosis, but it's also terrible to know!"*. In this group, patients were not afraid of being alone. Their annual check-up of the aorta diameter resulted in a feeling of safety. However, experiences of bodily pain and symptoms during exercise caused uncertainties, thinking: *"Is it my heart? My aorta?"*.

In three out of 11 participants, the level of psychological distress was prominent. These patients experienced a significant level of psychological distress on a daily basis. They described constantly being aware of the fact that they have an aortic aneurysm that could dissect or rupture. One participant said: *"It feels like having a 'time bomb', because you don't know when it goes off. And... you have to live with that knowledge"*. Two of the three were women; both diagnosed with a genetic predisposition for thoracic aortic aneurysm, positive family history of familial aneurysm and both already underwent surgery. The other patient was a man, without any known genetic disorder or familial aneurysm and with a thoracic aortic diameter of 50 mm. For these patients the diagnosis of the disease had been shocking, especially for the patient who had received surgery a short time after receiving the diagnosis. This patient expressed thoughts such as: *"Will the surgery be performed in time, what if the aorta will rupture before it's my turn?"*.

These feelings of prominent distress were triggered by bodily pain and symptoms, as expressed by one patient: *"I am feeling pain, does this mean there is something wrong with my aorta?"*. Two patients did not feel comfortable being alone, because they were afraid that something might happen. One of the patients, experienced distress especially when family members expressed their fear. In one participant religion played an important role, thinking: *"Do I take good care of my 'borrowed' body?"*. While another patient who experienced moderate psychological distress, felt supported by her religion.

### Communication

Communication with their healthcare team had little influence on the level of distress expressed by patients. However, when patients experienced having a 'positive' conversation with the physician, during which confirmation was given that the diameter of the aorta remained stable, this was reassuring. Receiving additional information by the physician about the disease and prognosis, was very much appreciated.

The communication within the familial context appeared to have a bigger influence on the level of psychological distress as compared to communication with the healthcare team. Eight out of 11 participants had an 'open' communication style about their disease with family members. In these cases, patients could freely talk about their worries with family members, which resulted in decreased feelings of distress. However, such an 'open' communication climate in the family was also found to be stressful for the whole family. For example, when posing the question if there is any kind of fear having this aorta disease, a family member replied: *"Do they really think we don't have any fear, of course we have fear!"*. Three participants did not talk about their disease with their family members, as they deliberately did not want them to worry about their condition. Although most patients had a network of friends whom they could talk to about their disease, these conversations were experienced as superficial by patients.

### Sense of control

#### Disease-related knowledge

Patients described that having more knowledge about their disease could give them a stronger feeling of control resulting in decreased levels of distress. On the one hand, a lack of disease-related knowledge of the participant or the family physician, gave a loss of control resulting in an increased level of distress. Some participants self-fulfilled this condition and increased their level of disease-related knowledge by reading about the disease, attending scientific meetings or participating in a patient association. On the other hand, some patients described that 'having more knowledge about the disease' resulted into more awareness and subsequently generated more distress.

#### Unpredictable and uncertain prognosis

Almost all participants reported that the unpredictable and uncertain evolution of the disease triggered feelings of loss of control and thereby increased levels of psychological distress. Some participants said: *"They can't guarantee that the aorta will not rupture. If the aorta will rupture, than you will die at once"*. This uncertainty again plays a major role in the degree of distress expressed by patients. *"If something will happen, is there a chance that I can have surgery on time and that I can make it?"* Shortly before a scheduled check-up of the aorta diameter, all patients expressed increased feelings of distress, which mostly disappeared when they received confirmation that the diameter had remained stable .

### Coping strategies

Coping strategies employed by our sample can be divided in two categories: (i) related to how the individual (the participant) handles his/her disease and (ii) in relationship with their partner, family member(s) and environment.

#### Coping style in relation to the self

Two participants seemed to have an inadequate coping style, resulting in a daily state of emotional distress due to their altered future perspectives. They experienced problems because they can't live their life the way they had planned before being diagnosed with their aortic disease. They said:

*"Honestly, I don't feel good at all, I'm always tired, I can't sleep and at my work I forget everything". "I can't be myself like I was in the past".* Any proposal to talk about their problems with a healthcare professional was, however, refused because they believed nobody could take away the fear they experienced. Other participants eventually accepted their disease, although most of them first experienced a period of shock, anger and sadness, having to change their sport practice or profession. For example: *"Then you have to adjust yourself, I couldn't play soccer anymore, that was really a bummer".* Feelings of acceptance were also encountered by some patients, as expressed by this patient *"I can worry, but I live now and when I continue to worry, I may live shorter".* Some patients have a generally more positive attitude towards life because of their aortic disease, they felt like they are living in spare time. These patients were grateful that they could be operated in time, increasing their chances to live a longer life.

#### Coping style in relation to partner, family and environment

Coping with the disease in relation to the partner, family and environment was seen as something individual by the participants. Patients carry their own worries and uncertainties, as if it was their individual responsibility. Illustrated by this quote: *"I know it can be hereditary, but I don't want to burden my children with it", "Don't want to bother them in their fantasies, life and stress".* In contrast, sharing with family members made them take over some tasks to relieve the participant.

#### Feelings of depression

Two participants expressed depressive thoughts, although these patients experienced these feelings in a very different way. One patient expressed depressive complaints at the moment of the interview. In this patient, the depressive thoughts were partly related to the aorta disease, although not exclusively. Other factors were: physical complaints not related to the aorta; being confronted with a partner who was depressed; having no other family members to rely on; and the recent suicide of a family member. In this participant, a constructive coping strategy for his disease was totally lacking and he explicitly did not want any professional help for his emotional problems. The other participant, however, described experiencing depressive thoughts in the past, shortly after receiving aortic surgery and losing a family member. At the moment of the interview, these thoughts were not present anymore. Many family members of this patient were diagnosed with aortic pathology, underwent surgery and were confronted with death originating from the aortic disease. *"I know I was ehhe tired of life, when my third brother....than I thought, arrggh, I don't have the energy for it, for mourning, I don't want to anymore".* This patient sought professional help from a psychologist who helped her tackle these difficult circumstances and described that those negative thoughts disappeared largely. Meanwhile, this participant developed a very good coping strategy to live with her disease.

### Physical complaints

All participants reported experiencing some physical complaints, like thoracic pain, dyspnea or tachycardia. These complaints often triggered increased levels of distress. Most patients described an unsettling feeling when having physical complaints, because they wondered whether these complaints could be directly linked to their aortic disease or the heart.

### Sense of loss

The aortic disease had an impact on the patient's daily life, their spare time and social life. Furthermore, the majority of patients reported that the diagnosis itself had a significant impact on their personality, their professional choice and future, and thoughts about family planning. *"Yes, that's though, you suddenly have to adjust yourself, I wasn't allowed to play soccer anymore. I had many friends there, which I lost, because I stopped"*. Another participant could not do his job anymore, because he was not allowed to lift weights. One participant felt angry towards life when she had to work fewer hours, and have less responsibility. *"The biggest problem of this disease is that I couldn't do the things that I have always done"*. Three of the 11 participants informed their sports instructor about their disease, in case something happened during exercise. This makes them less free to do sports anywhere, limited their personal space and freedom.

On the other hand, five participants reported living without limits and doing everything they wanted to do. They deliberately choose not to let their disease determine what they can or cannot do. The participants who underwent surgery in the past expressed a significant decline of their physical health condition, shortly after their operation, but also many years later. Three participants said they are not themselves anymore; they changed into a different personality due to the condition, being more emotional or easily triggered by certain events.

One participant even described that her diagnosis had a significant impact on her decision not to have children and start a family. However, two out of seven patients who had already children before they knew about their hereditary disease, did not know what they would have done if they knew about their diagnosis beforehand.