

LIVING WITH TRANSTHYRETIN AMYLOID CARDIOMYOPATHY: A TOOLKIT FOR PATIENTS



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AUTHOR SUPPORT AND ACKNOWLEDGMENTS

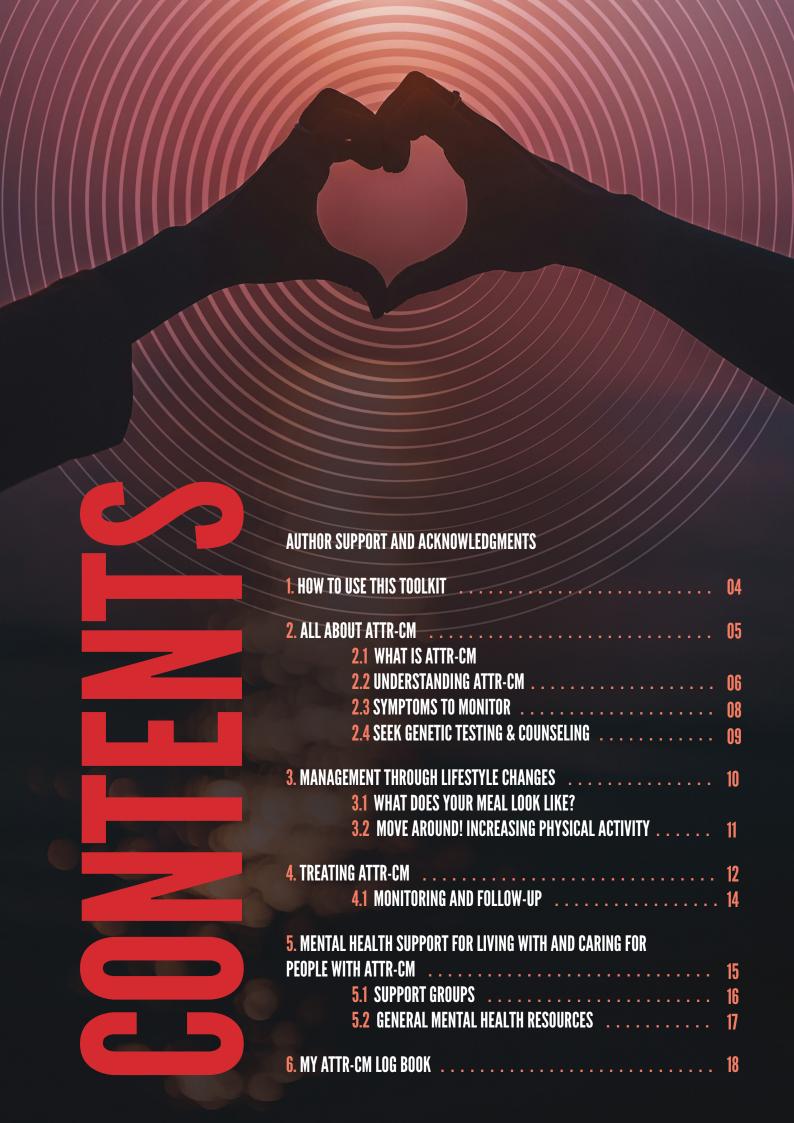
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THIS TOOLKIT HAS BEEN DEVELOPED BY WHF WITH SUPPORT FROM ALNYLAM, ASTRA ZENECA, PFIZER AND BRIDGEBIO.





1. HOW TO USE THIS TOOLKIT?

WHO IS THIS TOOLKIT FOR?

World Heart Federation has designed this simple toolkit to support people (both patients and carers) living with Transthyretin Amyloid Cardiomyopathy (ATTR-CM).

WHY HAVE WE MADE THIS TOOLKIT?

An ATTR-CM diagnosis can be challenging to navigate. The vast information available from multiple sources can be complicated and confusing to process. This document is designed to act as a supporting tool to help you better understand how to manage your symptoms and disease progression.

WHAT IS INCLUDED IN THIS TOOLKIT?

This toolkit includes education, lifestyle modifications, and resources to help manage the condition effectively. Patients should work closely with their healthcare team to personalize this toolkit to their specific needs.

WHAT IS UNIQUE ABOUT THIS TOOLKIT?

This toolkit is designed for YOU, the patient, and requires your input to monitor, track, and manage ATTR-CM symptoms. It does not contain a great deal of information on the science and medicine behind ATTR-CM but rather is meant to act as a pocket library of resources and tools to manage life with ATTR-CM.

HOW CAN YOU USE THESE TOOLS?

We have compiled several resources, including support groups and mental health resources for both patients and the carers of people with ATTR-CM. These can be accessed at will and tried and tested to find a support mechanism that best suits you. The 'My ATTR-CM Log Book' has been designed so a range of information can be easily accessed when needed and from a single place. It covers:

- Your diagnosis
- Your healthcare providers
- Your medications
- Your symptoms
- Your mental health and lifestyle.

2. ALL ABOUT ATTR-CM

2.1 WHAT IS ATTR-CM

Transthyretin Amyloid Cardiomyopathy (ATTR-CM) is caused when a protein (called transthyretin or TTR) that is found in the blood changes its shape. This change in shape makes it build up in the heart, nerves, and other organs. These build-ups are called amyloid deposits.

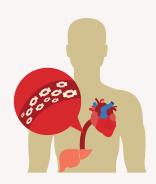
When the amyloid deposits are found in the heart, they make the muscles in the walls of the heart stiff, so the heart struggles to squeeze properly. This means that the heart is not able to pump as well as it should, and not enough blood is pumped around the heart and body. This condition is called cardiomyopathy and if left untreated, can ultimately lead to heart failure.

2.2 UNDERSTANDING ATTR-CM



1 NORMAL STATE

The liver produces transthyretin (TTR), a protein that helps transport vitamin A and thyroid hormones. In a healthy state, it remains stable.



2. AMYLOIDOSIS-MISFOLDED ATTR PROTEIN

In ATTR, TTR misfolds and forms amyloid deposits, accumulating over time and disrupting heart and nerve function.



3. BUILD UP OF ATTR DEPOSITS IN THE HEART

Misfolded TTR deposits in the heart, causing the walls to thicken and stiffen, reducing flexibility.



4. ATTR CARDIOMYOPATHY- HEART STRUGGLES TO FILL AND RELAX

A stiff heart has difficulty relaxing and filling with blood, leading to poor oxygen circulation and fatigue.



5. PROGRESSION TO HEART FAILURE

As ATTR progresses, the heart weakens, struggles to pump blood effectively, and causes fluid buildup, swelling, and extreme fatigue.

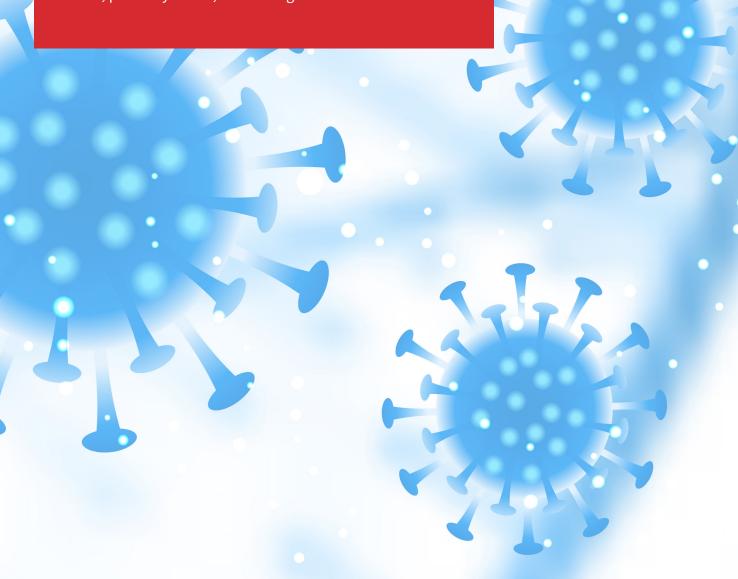
There are two different types of ATTR-CM:

HEREDITARY VARIANT:

This means that the changed protein gene is passed down through families, with symptoms being present earlier (as early as 30 years of age) than the other form of ATTR-CM.

WILD-TYPE:

This is more common than the hereditary variant form and does not run in families. The wild-type form of ATTR-CM is more commonly found in older individuals, primarily males, over the age of 60.

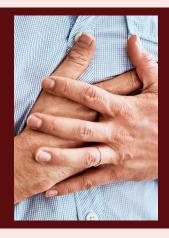


2.3 SYMPTOMS TO MONITOR

Both hereditary and wild-type ATTR-CM symptoms can really differ between patients, and are often subtle! Some people don't experience any symptoms at all until the late stages, when organ damage has already occurred.

This condition is very often misdiagnosed as the early symptoms are similar to those of other conditions, such as heart failure or high blood pressure.

The most common symptoms to monitor are:



SHORTNESS OF BREATH:

Especially when you walk or exercise, and even at rest in the later stages.



SWELLING IN THE LEGS OR ABDOMEN:

Legs or stomach feeling swollen and tight, such that if you press the area with your finger, it can leave an indent in your skin (this is more common in legs).



CHEST CONGESTION:

Coughing or wheezing.



FATIGUE:

Feeling tired even when you have had a good night's sleep.



IRREGULAR HEARTBEATS:

When you feel your heart beating at a different rate compared to normal, or even notice missing beats.



DIZZINESS OR LIGHT HEADEDNESS.

Feeling like the room is spinning, or that you are going to faint.

Contact your healthcare provider if you notice your symptoms worsening or you develop any new symptoms. Seek immediate medical attention if you feel like you are unable to breathe or have pain in your chest.

2.4 SEEK GENETIC TESTING AND COUNSELING

Since there are two types of ATTR, it is important to get a genetic test done to determine whether it is wild-type or hereditary. In the case it is hereditary, you should seek genetic counseling to identify relatives who might be at risk, so they can also contact their healthcare provider.



3. MANAGEMENT THROUGH LIFESTYLE CHANGES

There are several ways to manage or reduce the severity of the symptoms through implementing some lifestyle changes.

3.1 WHAT DOES YOUR MEAL LOOK LIKE?

- TRY AND FOLLOW A HEART-HEALTHY DIET THAT IS:
 - **Low** in saturated fats like butter, cheese, cream, chocolate, cakes, fried foods, and oils.
 - ▶ High in vegetables, fruits, whole grains, and lean proteins - non-fatty meats, chicken, fish, tofu, soya beans, chickpeas, and lentils.
 - Low in sodium. Try and keep salt intake to a minimum, as this will help reduce fluid retention and swelling.
 - Try and reduce the amount of sugar consumed, both through eating and drinking. Limit eating things such as pastries, cakes, chocolates, and sweets every day. Swap full sugar carbonated or juice drinks for sugarfree as well as swapping sugar in drinks like coffee or tea for sweetener.
- HAVE SMALL MEALS BUT EAT SEVERAL TIMES A DAY.
- MONITOR HOW MUCH LIQUID YOU DRINK DAILY AGAINST HOW MUCH IS PASSED AS URINE, ESPECIALLY FOR THOSE WHO EXPERIENCE SWELLING.



3.2 MOVE AROUND! INCREASE PHYSICAL ACTIVITY



REMEMBER THAT

things like gardening, cooking, and household tasks are forms of exercise too.





KEEP MOVING!

Try and include lowimpact exercise into daily life, such as walking, swimming, and bike riding if you can.





ONLY DO WHAT YOU CAN DO AND PACE

YOURSELF. Be aware of your own limitations and your body without overdoing things and making symptoms worse. If you start to feel tired while doing any form of physical activity, it is important to take a break or completely stop and rest. It is better to do little and often, rather than a lot followed by nothing for days because you are too fatigued or unwell to get out of bed.





4. TREATING ATTR-CM

There is no cure for ATTR-CM yet and the number of medications approved to slow down the progression of ATTR-CM varies by country and is updated as new treatment options become available. Please check with your healthcare provider for the latest status in your country of residence.

BEFORE TAKING ANY MEDICATION,
IT IS VERY IMPORTANT TO LET YOUR
HEALTHCARE PROVIDER KNOW OF ANY
EXISTING MEDICAL ISSUES YOU MAY
HAVE, WHETHER YOU ARE OR COULD
POSSIBLY BE PREGNANT OR IF YOU ARE
OR PLAN TO BREASTFEED WHILST ON
MEDICATIONS.

IT IS ALSO VERY IMPORTANT TO TELL THEM OF ALL THE

- MEDICATIONS,
- OVER THE COUNTER MEDICINES,
- HERBAL MEDICINES,
- SUPPLEMENTS

YOU ARE CURRENTLY TAKING AS SOME MEDICATIONS OR SUPPLEMENTS MAY NOT WORK WELL TOGETHER.

Some medications can slow down the disease progression by halting or reducing the build-up of protein deposits. Examples of these drug types and classes have been given below. Please consult with your physician for any questions and prior to starting any medication.



KEEPING UP WITH YOUR PRESCRIBED MEDICATION:

It is very important that you are regular with taking all the medications you have been prescribed on time. You might want to consider using medication tracking apps to remind you or even use a daily/weekly pillbox to help you organize the medicines.

TTR STABILIZERS

Tafamidis and Acoramidis are TTR Stabilizers. These medications do not break down protein build-ups that are already there BUT they do stop the build-up getting worse by preventing the proteins from changing shape.

This means it is important to start this treatment early before the disease has had a chance to do too much damage. As with all medications, it is important to take them as they are prescribed.

OTHER THERAPIES BEING STUDIED OR USED FOR SYMPTOM MANAGEMENT:

Some medications, known as TTR silencers, slow down the liver's production of faulty amyloid proteins and are approved for use in patients with hereditary ATTR-associated polyneuropathy. They are currently being studied to see whether they can be used to treat ATTR-CM.

Some other treatment options focus on managing the symptoms and trying to improve the quality of life. These include:

- Reducing dietary sodium intake (e.g. through a low-sodium diet)
- Diuretic medication
- Anti-arrhythmic medication to regulate the irregular heart rate.



4.1 MONITORING AND FOLLOW-UP

Keeping in regular contact with healthcare providers and cardiologists is **VERY** so that they can monitor your symptoms. It is a good idea to monitor your blood pressure at home on a regular basis and keep a log of the readings so you can show them to your healthcare providers when you see them. You can get blood pressure monitors to use at home quite cheaply. It is best to use one that has an arm cuff that is like the ones that are used in a clinical setting (with the large cuffs). They give more accurate reading than phone apps.

There is a blood pressure, heart rate and blood oxygen level (SpO₂) monitoring sheet at the back of this booklet you might find useful. It is also good to just use a regular notebook or writing pad to write down the readings as well. Alternatively, you can also use an activity tracker or smart watch with a built-in activity tracker to monitor your heart rate, SpO₂, etc. Find what works best for you!

Remember to state the date and time when the readings were taken and make sure you are sitting down quietly and not talking when taking the readings. This is especially important for the blood pressure readings.



STAY PREPARED FOR EMERGENCIES!

Put together a simple action plan for emergencies and when you or a caregiver should seek urgent medical care for you. For example, if you experience severe shortness of breath, chest pain or fainting.



EMERGENCY CONTACTS

In case of emergency, keep a list of emergency contacts including healthcare providers, family and friends. Having these at hand in one place can be helpful if needed in a hurry.

5. MENTAL HEALTH SUPPORT FOR LIVING WITH AND CARING FOR PEOPLE WITH ATTR-CM

An ATTR-CM diagnosis can be mentally taxing on not only the patient, but also on close family, friends and carers. It is very important to look after your mental health so you can pay attention to your physical health!



5.1 SUPPORT GROUPS

It can be a very scary and lonely time when you are diagnosed with a heart condition. Joining a support group may help you feel less scared and alone knowing there are others going through similar things as you.

There will be local groups you can access and go to in your local community, as well as online groups where other people can offer advice, support, share experiences and coping strategies. These safe spaces are also a good place to talk about your fears and worries, as you will not be the only one experiencing these emotions. It can be hard talking and opening up to family members or people close to you as you may feel the need to be strong for them.

Remember, it is ok to be not ok, and support groups can allow you to express and explore your true feelings and emotions with others that understand.

Here are a few resources that may be useful:

International: https://www.amyloidosisalliance.org/

Australia: https://aan.org.au/health-professionals/amyloidosis-types/attr/

Belgium: https://www.amybel.be/

Brazil: www.abpar.org.br

Canada: www.amyloidosiscanada.org

France: www.amylose.asso.fr

 $\textbf{Germany:}\ \underline{www.amyloidose\text{-}selbsthilfe.de}$

Netherlands: <u>www.amyloidose.nl</u> **Sweden**: <u>www.famynorrbotten.se</u>

United Kingdom: www.amyloidosisuk.org

United States: www.amyloidosis.org / https://www.amyloidosissupport.org/

There are also several public and closed Facebook groups and forums to connect with other individuals going through a similar experience including:

Amyloidosis Research Consortium/ Amyloidosis Research Consortium (ARC)

<u>Amyloidosis International</u>

Amyloidosis FAP-TTR-Hereditary

<u>Amyloidosis – Wild-type – ATTR</u>

Amyloidosis Awareness

It can also be a difficult time for your family and those who care about you. Here are a few resources that may be of support for them as caregivers:

Family Caregiver Alliance: www.caregiver.org

National Alliance for Caregivers: www.caregiving.org

Caregiver Action Network: www.caregiveraction.org

Carers UK: www.carersuk.org

Caringbridge: www.caringbridge.org/

Carers Australia: www.carersaustralia.com.au

Caregivers France: www.lappui.org
Caregivers Greece: https://epioni.gr/



5.2 GENERAL MENTAL HEALTH RESOURCES

It is also important to look after your own mental health. Consider practicing relaxation techniques, breathing exercises or yoga to help manage any stress and improve your overall mental health. There are also several mobile apps available for mental health and wellness. If you are comfortable using these, explore and see what suits you best. Being diagnosed with a chronic condition can be very difficult to come to terms with and it is not unusual to have feelings of anxiety or depression. If this is the case, then please think about seeking therapy or counseling to help work through these feelings.

Below are some resources for general information about mental health and support.

- Mind UK: www.mind.org.uk
- Mental Health America: www.mhanational.org







MY ATTR-CM LOGBOOK



NAME YEAR

6.1 WHAT IS IN THIS LOG-BOOK?

1. IN CASE OF EMERGENCY

A current list of medications, dosage, contact numbers of family members or caregivers and medical professionals who are overseeing your care. A brief outline of when to call for medical assistance if you are too unwell to do so yourself.

2. SYMPTOM TRACKER

Monitoring Symptoms, Blood Pressure and Heart Rate Tracking (regular monitoring, prepares for appointments) (tracking changes helping to identify trends or triggers; comparison over time).

3. MEDICATION MONITORING SHEET

Medication Management (if and when medication is taken, side effects)

4. WELLNESS LOG

Lifestyle Habits (Diet and Physical Activity, Fluid Intake, Weight)

5. PREPARING FOR DOCTOR VISITS SHEET

Outlining questions that are important to you to be answered.



6. MY EXPERIENCES JOURNAL

Supporting emotional and psychological well-being and engaging family members and caregivers.

6.2 IN CASE OF EMERGENCY (ICE FORM)

EMERGENCY CONTACT DETAILS

- MY NAME
- MY ADDRESS
- MY PHONE NUMBER

ICE CONTACT NAME

ICE CONTACT ADDRESS

ICE CONTACT PHONE NUMBER

EMERGENCY CARE PLAN

If I am experiencing severe shortness of breath, chest pain or I faint and cannot call for medical help myself...

Our example is: please call for an ambulance and wait with me until they arrive.

ORGAN DONOR STATUS	BLOOD TYPE
ALLERGIES	

HEALTH CARE PROVIDERS INVOLVED IN MY CARE		
•		
DOCTOR		
PHONE NO.		
•		
CARDIOLOGIST		
PHONE NO.		
•		
NURSE		
PHONE NO.		
•		
CARER		
PHONE NO.		

CURRENT MEDICATION AND DOSES			
MEDICATION	DOSE		
1.			
2.			
3.			
4.			
5.			

IMPORTANT NOTES	

6.3 SYMPTOM TRACKER

ATTR-CM DAILY SYMPTOMS TRACKER - ABC FORM

(With thanks to INOCA International for the use of this tracker – please feel free to customize this tracker to be specific to your own symptoms etc)

Date / Time Location

29.06.20 12.35

What happened right before the symptoms began (Antecedent)

A = at rest 1 during the day, 2 woken from sleep

B = post exertion **1** walking, **2** showering, **3** drying hair,

4 housework, 5 other – please specify)

C = Environmental pollutants 1 noise, 2 lighting, 3 crowded environment, 4 other - please specify)

D = emotional stress or/situation

E = change in weather conditions **1** hot to cold, **2** cold to hot,

3 stormy, 4 other -please specify

F = other

G = unknown

F - zoom meeting at work/lounge

A1, B2, C1,

C2

Description of symptoms (Behavior)

 ${\bf A}$ = Heart Related ${\bf 1}$ increased heart rate ${\bf 2}$ irregular beats,

3 palpitations, 4 other - please specify

B = Breath related – especially when lying down/resting

1 coughing, 2 wheezing, 3 shortness of breath wheezing,

4 other - please specify

C = Trouble thinking **1** confusion, **2** word finding difficulty,

3 other - please specify

D = Swelling 1 tstomach/abdomen, 2 hands, 3 feet, 4 legs,

5 other please specify

E = exhaustion **1** cannot keep eyes open – have to sleep, **2** cannot

perform usual functions, **3** tired even after a good night's sleep,

4 other -please specify

В

How long symptoms are lasting

A = less than 1 minute

B = lasting 1-10 minutes

C = 10-30 minutes

D = 30+ minutes

E = unknown

What helped calm the symptoms (Consequence)

A = Deep breaths

B = Rest

C = Meditation

D = Rescue medication taken

please specify

E = Healthcare provider contacted for advice

F = Hospital emergency visit

G = Ambulance called

H = Other - please specify

A, C,

Observations BP/O2 etc

SO2 92% BP 138/87

SYMPTOM TRACKER

ATTR-CM DAILY SYMPTOMS TRACKER - ABC FORM

Date / Time Location			
What happened right before the symptoms began (Antecedent)			
Description of symptoms (Behavior)			
How long symptoms are lasting			
What helped calm the symptoms (Consequence)			
Observations BP/02 etc			

6.4 TIPS ON FILLING IN THE ABC FORM

FIRST

add the date and time in the bottom left hand white box.

THEN

move to the next white box to the right (under 'what happened right before the symptoms began').

LOOK

in the pale colored boxes to see which descriptions best fit your symptoms, for example, - if your symptoms came on at rest, during the day you would write 'A1' in the white box at the bottom of that column - with a description if you wish.

WORK

across the columns in the same way, matching your symptoms as you go.

ONCE

you have completed the 4 columns you will then have a row of information at the bottom, from left to right, that details your symptoms that day.

Completing the tracker each day may help you to identify a pattern or triggers that are personal to you. In our example we have color coded each section to show which section relates to which part in the below statement –

On 29th June at 12.35

I was in a video meeting (F1) in the lounge when I experienced increased heart rate (A1), wheezing (B2), confusion (C1) and word finding issues (C2). The symptoms lasted approximately 6 minutes (B). I took deep breaths (A) and meditated (C) which eased the symptoms). My oxygen levels and BP were SO2 92% BP 138/80.

6.5 CURRENT MEDICATION LIST

CURRENT MEDICATION	DOSE	SIDE EFFECTS EXPERIENCED	DISCUSSED WITH HEALTHCARE PROVIDER



6.6 WELLNESS LOG

DATE	SUN	MON	TUE	WED	THU	FRI	SAT
SLEEP QUALITY HOURS	& &	&&&	&&&	888	&&&	&&&	&&&
OF SLEEP	- 56789+	- 56789+	- 56789+	- 56789+	- 56789+	- 56789+	- 56789+
MEDICATIONS	000	888	888	888	888	888	888
BLOOD PRESSURE	(<u>*</u>	☼ •	₩	☼	₩	☼ •	☼
	ĕ.		₩	· i		₩.	
HEART RATE							(C)
SO2	©.	☼ ·	©.	☼ •	₩	☼ •	©.
5 DAILY	* *						
PORTIONS OF FRUIT AND VEGETABLES							
	Run Run Run	gue gue gue			gre gre gre		
8 GLASSES OF WATER A DAY							
8 GLASSES OF WATER A DAY							
8 GLASSES OF WATER							
8 GLASSES OF WATER A DAY MOVED							
8 GLASSES OF WATER A DAY MOVED MORE	\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\\$\\	\$\times\$					
8 GLASSES OF WATER A DAY MOVED MORE TIME FOR ME		• •					



6.7 PREPARING TO VISIT THE DOCTOR



DATE OF APPOINTMENT

DRS NAME

TICK THE BOXES BELOW TO INDICATE THE MODULES YOU PLAN TO DISCUSS WITH YOUR DOCTOR.







QUESTIONS	ANSWERS

6.8 MY EXPERIENCE JOURNAL

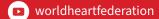
DAY/ DATE	HOW I'M FEELING AND WHY	WHAT NEXT
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