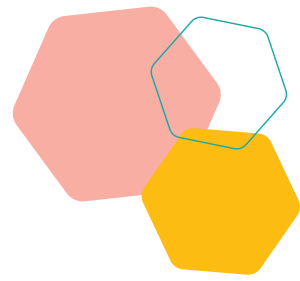


My SMA Diary



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Foreword

Spinal Muscular Atrophy (SMA) is a rare neuromuscular disease. Because it is rare, most doctors do not know SMA well.

An SMA diagnosis can be overwhelming, and I hope this My SMA Diary helps give persons with SMA and their families information about what kind of medical treatment they may receive in an accessible format, suitable for people who do not have a medical background. MY SMA Diary is therefore designed to help in observing and following a medical care-giving plan for SMA, which is important to contribute to the overall well-being of families with a child or adult who has SMA.

The guidance was developed by Persatuan WeCareJourney based on international consensus statements of standards of care in SMA, and the recommendations of top specialists who support SMA families in Malaysia. We would like to thank the doctors, patients with SMA and caregivers who have contributed their time to provide input and feedback in creating MY SMA Diary and also the generosity of our donors, sponsors and supporters, the experts and volunteers who have given their time, and our team. Terima kasih banyak banyak!

*Mr Edmund Lim
President of Persatuan WeCareJourney
And Spinal Muscular Atrophy and Disability
Activist and Advocate*



A Message from Dr Amar

Dear parents,

When your child is found to have Spinal Muscular Atrophy it will come as a major shock and challenge. There is no easy way to deal with this and each of us will respond differently. Remember not to blame or place guilt on yourself – this is not your fault and it has not happened because you did something wrong.

The key to moving forward – to helping your child and family – is to network with and meet other parents to share ideas, learn and get emotional support – this is vital. There will be many different specialists and therapists to see; if possible, find one person, whether a paediatrician, therapist or experienced NGO liaison, who can serve as a key coordinator to overview and plan therapy needs for your child.

I would recommend that both parents get involved and not allow the burden to be solely placed on the mother. Both parents need to down-size work and spend more time at home with their child and family. Remember that all members of the family will have needs, including the other children and the parents. Restructure the family to make more time for everyone in order to prevent long-term problems. Also make regular time for yourself to reflect and be with God.

Enjoy the small moments with the family to find joy – there is hope.

Much blessings to you,
Amar

*Dato' Dr Amar-Singh HSS
Senior Consultant Paediatrician
Ipoh, Perak, Malaysia*



Message from A Mother

Dear parents and carers,

Like you, I have a child with Spinal Muscular Atrophy (SMA). My son, Branden, was diagnosed with SMA when he was one year old. The news was shocking to my husband and I; we were lost, overwhelmed and didn't know what to do at all. We felt like we had just fallen into a cave. Even the word "SMA" was foreign to us.

The doctors gave us so much information about SMA, and what our child and family would have to go through. But it was hard to take it all in at once. At that time I wished I had someone or something to guide me.

Finding out that your child has SMA will be a new chapter in your family's life. It will be a new journey.

This booklet, MY SMA Diary, can help to be your "compass" in this new journey. It will guide you on how to manage and care for your child with SMA. When you use this Diary, you will also be able to monitor your child's progress, growth and development. It will also guide you in asking questions to the doctors involved in your child's care.

It is important to remember that you are not alone in this journey, you can reach out to other families in the SMA community for support. There will be challenges and tears, but there will be celebrations and laughter too.

Take care of yourself as well, as this is a long journey with Hope.



*Sook Yee, mother and Co Founder of
Persatuan WeCareJourney*

Words from Community



Mother of Adra,
a child with type 1 SMA

“Avoid focusing on your child’s weakness. Focus on their strength”

Mother of Dhaffa Rifqi,
a child with type 1 SMA

“ Every child is a gift and as a parent/caregiver, we are responsible to provide the best care, affection and guidance. Not every person has the opportunity to give love to their special child. We are the chosen one, the one who are extra strong and resilient to go future challenges ”



Mother of Jayson,
a child with type 3 SMA

“I know how you are feeling, I understand you. Diagnosis is not the end, we are in this together for the treatments and hope”



Sha Roose,
an adult with type 3 SMA

“This disease is no longer the end of the world, remember to reach out and ask many questions as possible. The community will help where they can. You’re not alone”



Acknowledgements

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How to Use This Guide

This book is made specifically for parents of children with Spinal Muscular Atrophy (SMA) and to be used by healthcare providers to help fill in vital information during the child's follow-up and visits to the hospital. There are two parts to this book. Part 1 is a comprehensive guide, consisting of important information about SMA and Part 2 is a record of medical information and assessment of your child. A Malay translation is provided for Part 1 of this book only.

As your child has SMA, you are being provided with this book to help you learn more about this disease and help your child with his / her journey. The diagnosis may be overwhelming, but we are here to assure and help you through this journey.



If you are a doctor / healthcare provider:

- Part 1 provides a simple, concise management plan for children with SMA.
- Kindly help to fill up reviews and the current condition of the child in Part 2 where applicable.
- Write down any new diagnosis, tests / procedures and new medications or treatments.
- Fill up the date and time for the next follow-up appointment.



If you are a parent:

- You can get more information on SMA in part 1.
- Bring this book during all follow-ups and hospital visits.
- Before your appointment, know the purpose of the visit and write down the questions for what you want to know.
- Bring someone to accompany you as they can help you ask questions and remember the doctor's advice.
- If a new medicine or treatment is prescribed, ask the doctor how it will help your child, what are the side effects, and if the problem can be treated in other ways.
- If a test or procedure is required, ask the doctor for the reason it needs to be done and the result.
- Know what to expect if you do not take the medicine or undergo the test / procedure.
- Check the next appointment date before you leave the clinic.
- Know how you can contact your doctor if you have any questions.

ABOUT MYSELF



PATIENT'S INFORMATION

Full name : _____

Date of birth : _____

Gender : _____

Race : _____

MyKid no : _____

Home address : _____



PARENT'S INFORMATION

Father's name : _____

Contact number : _____

IC no : _____

Occupation : _____

Email : _____

Mother's name : _____

Contact number : _____

IC no : _____

Occupation : _____

Email : _____



EMERGENCY CONTACT'S INFORMATION

Name : _____

Relationship : _____

Contact number : _____

Name : _____

Relationship : _____

Contact number : _____



SIBLINGS INFORMATION

No	Year of birth	Gender	Health Condition
1			
2			
3			
4			
5			

PART 1

INFORMATIVE GUIDE

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1.1 Introduction to SMA

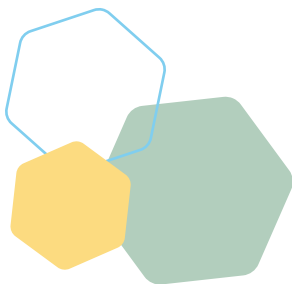
1.1.1 What is SMA?

SMA is a genetic disease. It affects the motor neuron cells in the spinal cord. The function of the cells is to send signals to the muscles to work, but in SMA, they do not function the same way, causing the muscles to become very weak.

People with SMA may have difficulty walking, eating, and even breathing, because of muscle weakness. Even though they have limited physical function, they are not impaired intellectually. Children with SMA can think, learn and build relationships with other people.

1.1.2 What causes SMA?

SMA is an inherited disease – one that is passed from parents to children. It is caused by a missing or faulty gene, i.e. SMN1 (survival motor neuron 1). This gene is responsible for the production of the survival motor neuron (SMN) protein that is critical for nerve function and control.



1.1.3 Four types of SMA

<p>TYPE 1 (Severe)</p> <p>Age of onset is less than six months.</p> <ul style="list-style-type: none">• Most severe form.• Within the first few months of birth, the baby may have severe muscle weakness, poor swallowing reflexes and difficulty breathing.• 95% of these children will not live past their second birthday unless they receive disease-modifying treatment. However, this is not currently available widely.	<p>TYPE 2 (Intermediate)</p> <ul style="list-style-type: none">• Age of onset is seven to 18 months.• Delay or failure in meeting motor milestones.• Can sit up (although might require assistance), but unable to walk, and requires a wheelchair.• Life expectancy may be shortened.
<p>TYPE 3 (Mild)</p> <ul style="list-style-type: none">• Age on onset is 18 months onwards• Children able to stand and walk initially, but will grow weaker, and many eventually need to use a wheelchair.• Chewing and swallowing may be affected later.• Life expectancy is generally normal.	<p>TYPE 4 (Adult-onset)</p> <ul style="list-style-type: none">• Age of onset is mid-30s onwards.• Late onset and similar to Type 3, but less severe.• Life expectancy is normal.

1.1.4 Which doctors do I see now?

SMA is a disease that can affect a few different systems in our body, such as the respiratory, neuromuscular and gastrointestinal systems. The healthcare team should comprise specialists/experts from neurology, genetics, respiratory care, rehabilitation, orthopaedics and nutrition. However, this is only available in big medical centres / hospitals. This will be discussed further in the following chapters. Remember that early treatment offers the best chance for a child to stay as healthy as possible.

While you may need to see many specialists and therapists, it would be good to have one doctor who acts as the family's main healthcare provider or coordinator, and looks at the overall needs for your child.

1.1.5 What to expect after diagnosis?

SMA varies from individual to individual. This is important to remember when considering different aspects of care. No two individuals will be the same. Treatment and care plans should be tailored to meet individual needs. This can be achieved with an interdisciplinary care team at a specialized care centre. It is also important to remember that SMA does not affect one's ability to think, feel and learn. Therefore, individuals with SMA have normal cognitive abilities. Participation in as many age - and developmentally-appropriate activities as possible is encouraged, with adaptations made wherever necessary.

1.1.6 What is the treatment for SMA?

There is currently no known cure for SMA. Treatments are given to manage symptoms and prevent complications. Current SMN-enhancing treatments for SMA approved by the U.S. Food and Drug Administration (FDA) include:

- **Evrystdi (risdiplam)** is a small molecule taken daily by mouth or through a g-tube that causes the SMN2 gene to make more complete SMN proteins.
- **Spinraza (nusinersen)** is an intra-thecal injection that targets the SMN2 gene, causing it to make more complete proteins.
- **Zolgensma (onasemnogene abeparvovec-xioi)** is a gene therapy that provides complementary and independent DNA to replace the function of the missing or mutated SMN1 gene.

Current SMN-enhancing treatments for SMA are not widely or easily available worldwide. Talk to your healthcare provider about current options available in Malaysia.

1.1.7 How do I take care of my child?

A child with SMA requires at-home care. It is vital that a child is comfortable and adequate adjustments are made at home to ensure that your child is growing happily and healthily. Proper at-home care can prevent frequent hospitalisations and emergency visits.

1.1.8 Are there any support groups available?

Yes. You do not have to do this alone. Reach out for help when you feel overwhelmed. There are many support groups / help available to you. We will provide this information at the end of the book. Support groups are very useful, but having one or two specific parents who have already made the journey with a child who has SMA, or who have already gone through the stage you are experiencing now, to talk to regularly is very helpful.

1.2 SMA – A Guide for Healthcare Providers



Diagnosis

- Child with hypotonia / motor delay
- SMA genetic testing through blood sample and referral to a neuromuscular specialist / geneticist / paediatrician



Paediatric

- Ensure pneumococcal and annual influenza vaccination
- Monitor disease progression
- Assess for deformities and contractures, spine for scoliosis, and hip
- Evaluate functioning / activities of daily living



Genetic

- Provide genetic counselling after diagnosis



Obstetrics & Gynaecology

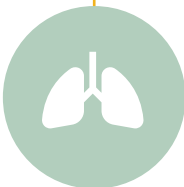
- Discuss prenatal screening for future pregnancies
- Offer amniocentesis and / or chorionic villus sampling for early detection

1.2 SMA – A Guide for Healthcare Providers



Rehabilitation & Orthopaedics

- Discuss and encourage contracture prevention with usage of splints / orthoses and frequent stretching
- Appropriate exercise
- Assistive mobility devices – strollers / scooters / wheelchairs
- Support airway clearance – oral suction/ physiotherapy / chest percussion / cough assist machine / postural drainage
- Additional examination (motor measures)
- Surgery by orthopaedics



Respiratory

- Assess for work of breathing and any sleep difficulties
- Nebulized bronchodilators for patient with asthma / positive bronchodilator response
- Chest X-ray when indicated
- Lung function test to record FEV1, FVC and TLC
- Peak cough flow test
- Sleep study to be performed or overnight pulse oximetry
- Bilevel non-invasive ventilation if indicated

1.2 SMA – A Guide for Healthcare Providers



Therapy

- Occupational therapy focusing on feeding, dressing and other activities of daily living
- Physiotherapy to improve mobility and exercises to maintain range of motion and improve function
- Continue daily stretching routines and use recommended braces or corrective devices (also known as orthotic devices)
- Speech therapy assessment if having difficulty in swallowing
- Support airway clearance – oral suction / physiotherapy / chest percussion / cough assist machine / postural drainage



Nutrition

- Monitor weight and height
- Assess / discuss diet
- Evaluate feeding and swallowing
- Treat gastroesophageal reflux and constipation as necessary
- Feeding tube when unable to meet all nutrient needs by mouth
- Tube feeding / gastrostomy, either bolus or continuous

1.2 SMA – A Guide for Healthcare Providers



Palliative

- Thorough approach on physical, psychosocial, emotional and spiritual issues to support patient and family members
- Discuss care during diagnosis, crises and terminal stage
- Advanced care plan when necessary

1.3 Genetics of SMA

Why do you need to see a geneticist?

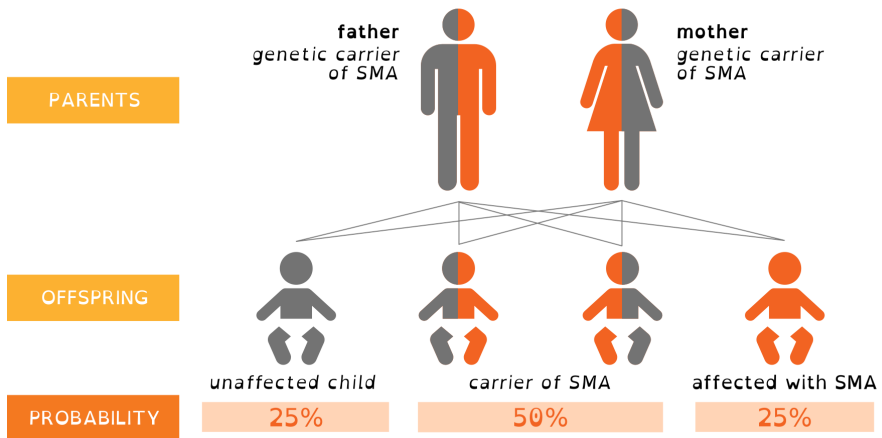
SMA is a genetic disease. You may have questions of how it is passed down in families and how you can prevent this disease in your future children. Here is a basic explanation about the genetics of SMA to help you understand more before discussing the topic with your doctor.

To better understand the genetics of SMA, here are a few key words and their meanings.

DNA: This is a molecule that contains the genetic code of living things. It links together in a sequence that makes up a gene.

Gene: This is a DNA sequence that will provide instructions for the body to make proteins.

Chromosome: This is a compact structure made up of DNA and protein.



Any error in a DNA sequence will affect the production of the protein the gene is responsible for. This error is known as a mutation.

In SMA, there is an error (mutation) in the SMN1 gene that produces the SMN protein. The SMN1 gene is found on chromosome 5 of the human genome. SMN proteins are critical to the function of the nerves that control muscle movement.

This mutation leads to motor neurons (nerve cells that send messages to and from muscles) not functioning normally and results in muscle weakness and atrophy (shrinkage).

It is estimated that one in 50 people throughout the world are carriers of SMA.

About one in 10 000 babies born are affected by SMA worldwide.

(About 50-60 babies born in Malaysia every year are affected by SMA)

How does one get diagnosed with SMA?

After an infant or child shows symptoms of SMA, a blood sample will be taken and sent for the SMN1 gene deletion test. If the test shows that both copies of the SMN1 gene have the deletion fault, a diagnosis of SMA will be given. It is recommended that the number of SMN2 copies are also counted as this can be a helpful indicator of how severely the condition will affect the child in the future.

Apart from diagnostic testing, there are other different types of genetic testing related to SMA.

Carrier testing : Confirms whether you are a carrier of SMA

Prenatal testing : Determines if your unborn baby has inherited SMA

Genetic testing is important as early diagnosis enables early treatment, which results in a better outcome.

It is important that family members understand the results of the genetic testing that led to the diagnosis of SMA. Genetic counselling should take place with a healthcare professional who has expert training in genetics.

If you plan on having more children, there are a few screening options available for parents who are known carriers of SMA.

1. **Chorionic villus sampling**

Performed as early as the 10th-13th week of pregnancy

2. **Amniocentesis**

Performed between 15th- 20th week of pregnancy

3. **Pre-implantation genetic diagnosis (PGD)**

Done before pregnancy

1.4 Respiratory Care

Why does your child need respiratory care?

A child with SMA can have underdeveloped lungs, a weak cough, increased infection rates, swallowing problems, aspiration, and breathing difficulties during sleep. Children with SMA need regular evaluations for their respiratory care.

What are the recommendations for my child?

Type of SMA	Intervention	Care Considerations
TYPE 1 Non-sitters <i>(unable to sit unsupported)</i>	Coughing (or airway clearance) is necessary to remove nasal secretions and inhaled particles. Coughing can be improved with <ul style="list-style-type: none"> • Oral suctioning • Chest physiotherapy • Manual chest percussion • Cough insufflatorex sufflator • Postural drainage 	
	If your child experiences breathing difficulties during sleep, he / she may need to use bilevel non-invasive ventilation (NIV).	This is initiated after observing the patient clinically for adequate gas exchange / sleep study
	Nebulised bronchodilators in patients with asthma / positive bronchodilator response.	Mucolytics should not be used long term.
	Patients also need yearly influenza and pneumococcal vaccinations.	

Type of SMA	Intervention	Care Considerations
<p>TYPE 2 / TYPE 3</p>	<p>Coughing (or airway clearance) is necessary to remove nasal secretions and inhaled particles. Coughing can be improved with</p> <ul style="list-style-type: none"> • Oral suctioning • Chest physiotherapy • Manual chest percussion • Cough insufflator-exsufflator • Postural drainage 	
<p>Sitters (able to sit independently)</p> <p>who become non-ambulant</p>	<p>If your child experiences breathing difficulties during sleep, bilevel non-invasive ventilation (NIV) may be considered.</p>	<p>Some experts recommend NIV during acute respiratory illness to facilitate discharge.</p> <p>This is also initiated after observing the patient clinically for adequate gas exchange / sleep study.</p>
	<p>Nebulised bronchodilators in patients suspected of having asthma.</p>	<p>Mucolytics should not be used long term.</p>
	<p>Patients also need yearly influenza and pneumococcal vaccinations.</p>	

Type of SMA	Intervention	Care Considerations
TYPE3/ TYPE4 Standers/ Walkers <i>(ambulant patient)</i>	Patients who are standers / walkers require less respiratory attention. Provide them with supportive care when needed.	Evidence of weak cough, recurrent infections or suspicion of nocturnal hypoventilation.
	Patients also need yearly influenza and pneumococcal vaccinations.	

Examples of devices used in intervention



Cough insufflator-exsufflator

Helps to clear secretions by applying a positive pressure to fill the lungs, then quickly switching to a negative pressure to produce a high expiratory flow rate and simulate a cough



Bilevel Positive Airway Pressure (BiPAP)

A type of non-invasive ventilation supplies pressurized air into your airways

1.5 Rehabilitation & Orthopaedics Care

Why does your child need rehabilitation and orthopaedics care?

A child with SMA may face limitations in range of motion of the joints, hip dislocation, spinal deformities (like scoliosis) and fractures. Changes in the musculoskeletal system, such as contractures, fractures and scoliosis, can lead to pain and difficulty with sitting and functional activities.

Example of orthoses



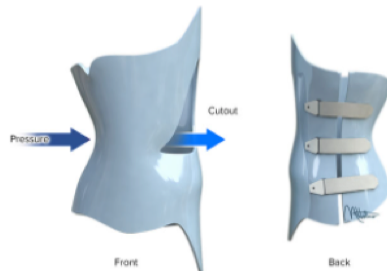
**Paediatric AFO
(Ankle Foot Orthosis)**



Soft Frame TLSO



Wrist Hand Orthosis-ORFIT



Boston Type Scoliosis TLSO

What are the suggested recommendations for my child?

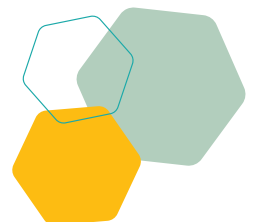
Type of SMA	Intervention	Care Considerations
<p>TYPE 1</p> <p>Non-sitters (unable to sit unsupported)</p>	<p>1) Positioning and bracing</p> <ul style="list-style-type: none"> • Use seating systems, postural and positioning supports • Use thoracic and cervical bracing for head support <p>2) Stretching</p> <ul style="list-style-type: none"> • Daily stretching activities should be done • Daily use of upper limb orthoses for stretching and to promote better function and range of motion • Static orthoses, knee immobilizers and hand splints for positioning and stretching • Wearing an ankle-foot orthosis (AFO) and knee-ankle-foot orthosis (KAFO) as much as possible will help maintain flexibility • Thoraco-lumbo-sacral orthosis • (TLSO) can be used for positioning • Try supported standing <p>3) Promote function and mobility</p> <ul style="list-style-type: none"> • Use of seating and mobility system 	<p>Orthoses to be used more than 60 minutes to overnight.</p> <p>Minimal frequency for stretching is three to five times per week.</p> <p>Bracing five times per week.</p>

What are the suggested recommendations for my child?

Type of SMA	Intervention	Care Considerations
<p>TYPE 2 / TYPE 3</p> <p>Sitters</p> <p><i>(able to sit independently)</i></p> <p>who become non-ambulant</p>	<p>1) Positioning and bracing</p> <ul style="list-style-type: none"> • Use thoracic and cervical bracing for posture and to promote function <p>2) Stretching</p> <ul style="list-style-type: none"> • Use upper and lower limb orthoses to help promote function and range of motion • Stretch areas at risk of contractures (e.g. hips, knees, ankles, wrists and hands) • Knee immobilisers, KAFO and AFO for position and standing • Reciprocal Gait Orthosis (RGO) and KAFO for supported ambulation • TLSO and hand splint for positioning <p>3) Promote function and mobility</p> <ul style="list-style-type: none"> • Use of seating and mobility devices • Use of gait training device and mobility device to support ambulation • Mobile arm supports to assist upper extremity function 	

What are the suggested recommendations for my child?

Type of SMA	Intervention	Care Considerations
<p>TYPE 3 / TYPE 4</p> <p>Standers/ Walkers</p> <p>(ambulant patient)</p>	<p>1) Promote function and mobility</p> <ul style="list-style-type: none"> • Recommend aerobics / general conditioning exercises <p>2) Stretching</p> <ul style="list-style-type: none"> • Active assisted stretching • Orthoses <p>3) Position and bracing</p> <ul style="list-style-type: none"> • Lower limb orthoses • Thoracic bracing 	<p>Exercise at least 30 minutes / day.</p> <p>Stretching should be done a minimum of two to three times per week. The optimal number is three to five times per week.</p>



1.6 Nutrition Care

Why does your child need nutrition care?

A child with SMA may face feeding and swallowing problems, gastrointestinal dysfunctions (e.g. constipation), delayed gastric emptying, gastroesophageal reflux, growth problem, undernutrition / overnutrition, and aspiration that can cause pneumonia

What are the suggested recommendations for my child?

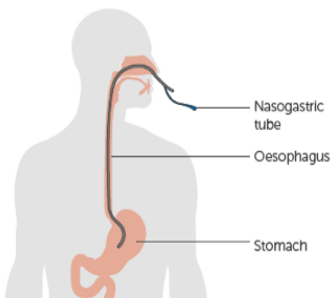
Referral to a dietician or paediatric gastroenterologist, who may recommend changes to your child's diet and other treatments, depending on individual needs.

Not all children will require a feeding tube. However, certain cases such as being unable to swallow safely, being unable to get all their nutrient needs by mouth, not growing or maintaining well, unable to drink enough fluids, ill, or before / after surgery, may require one.

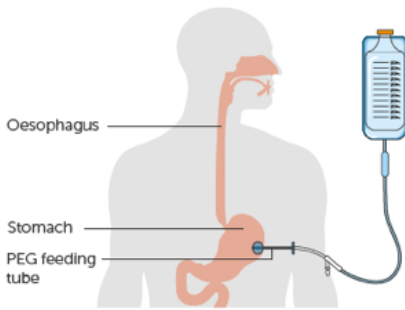
Types of tubes

Nasogastric (NG), Nasoduodenal (ND), Nasojejunal (NJ)

- Temporary or permanent
- Can be placed without surgery
- Complications: Sore throat, dislodgement, sinusitis

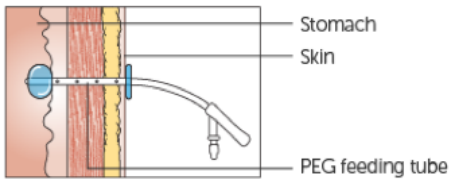


Percutaneous Gastrostomy (PEG) or Gastrostomy (G) Tube



- A long-term solution
- Placed surgically or through a scope
- Placed when indicated
- Complications: Infection, dislodgement

Gastrostomy-Jejunostomy (G-J) tube



- A long-term solution
- Placed surgically
- Placed when indicated
- Formula given at a slowerrate through the J-Port and requires a longer time on the feeding pump

After a tube is placed, a prescription for formula will be given. The most common tube feeding methods are **bolus** or **continuous feeding**.

1.7 Palliative Care

Why does your child need palliative care?

Every child and family with SMA type I can benefit from, and should receive, palliative care throughout their illness, regardless of the therapies that they choose. Palliative care helps maximize the quality of life for a child living with a serious illness such as SMA.

The term “palliative care” can be frightening or overwhelming for many families as it mistakenly and automatically makes people think only of hospice or end of life (EOL) care, and death. You will ideally be introduced to the concept of palliative care at the time of diagnosis, with this care being provided throughout the illness as decisions are made on an ongoing basis.

This is a brief overview of topics you might want to discuss with your healthcare provider

Physical condition of your child

- General condition and comfort level
- Respiratory system
- Musculoskeletal system
- Neurological system
- Gastrointestinal system
- Physical progress over time

Psychological well-being of patient and caregiver

- Level of understanding about the disease and phase of acceptance.
- Mental health of child with progression of age, as well as that of the caregiver.
- Family mental health, including siblings.

Social well-being for both patient and caregiver

- Interaction with people other than family members.
- Involvement in community and social support.
- Financial and accommodation challenges.

Spiritual well-being for both patient and caregiver

- Involvement in religious or spiritual beliefs.
- Support within religious or spiritual belief.
- Acceptance of life and death within own belief.
- Community involvement through religious or belief circle

Advanced care plan

1. Place of care
2. Wishes
3. Detailed plan involving respiratory tube and symptom / management.
4. Do Not Resuscitate (DNR) status
5. End of life planning

PART 2

ASSESSMENT RECORD

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2.2 Vaccination Schedule

Vaccine	Expected age to be given	Date Given
BCG	At birth	
Hepatitis B	At birth	
DTaP- IPV-Hep B- Hib	2 months old	Dose 1:
	3 months old	Dose 2:
	5 months old	Dose 3:
	18 months old	Booster:
Measles (Sabah & Orang Asli)	6 months old	
MMR	9 months old	Dose 1:
	12 months old	Dose 2:
MR	7 years old	
DT	7 years old	
HPV	13 years old	Dose 1:
	13 years old	Dose 2:
Tetanus	15 years old	
JE (Sarawak)	9 months old	Dose 1:
	21 months old	Dose 2:

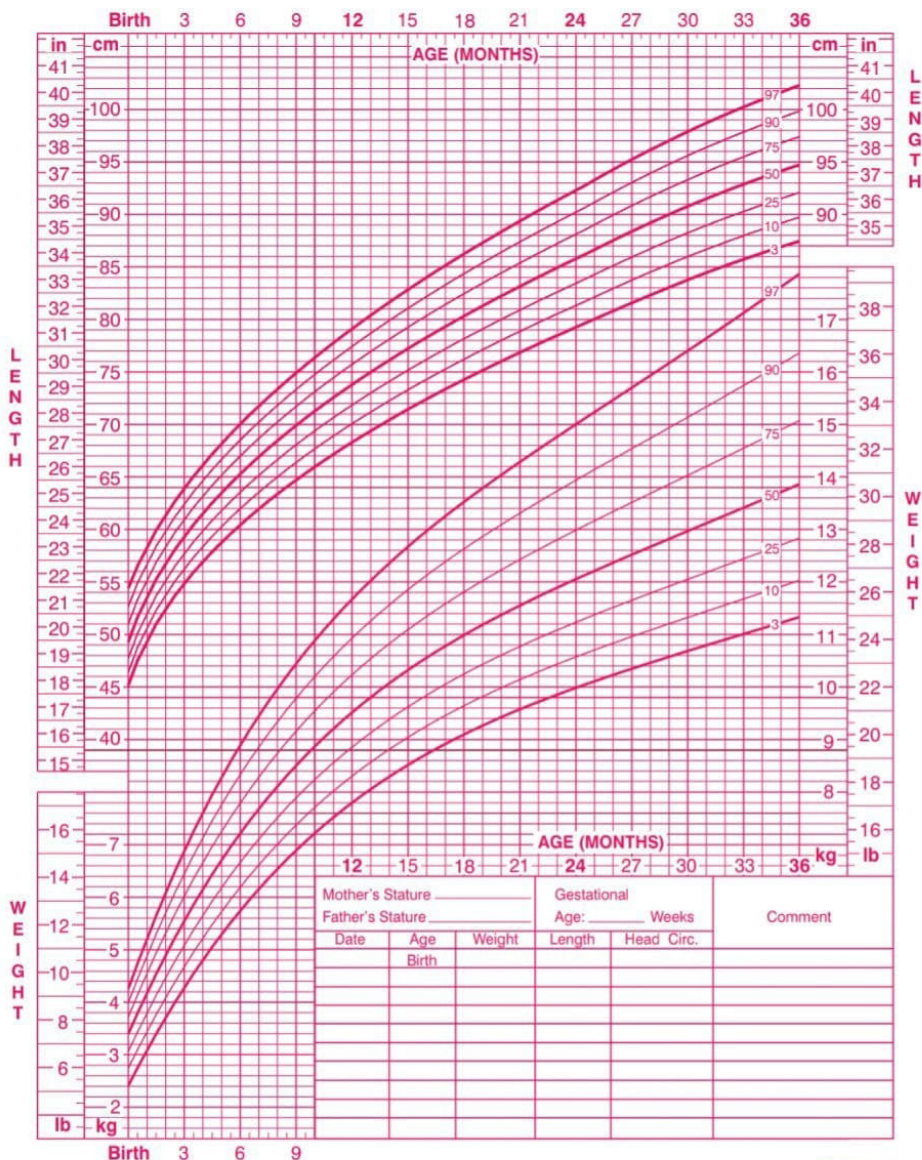
2.2 Vaccination Schedule

Vaccine	Expected age to be given	Date Given YY/MM/DD
Influenza (annually)		
	Pneumococcus	
		Dose 2:
		Booster:

Birth to 36 months: Girls
Length-for-age and Weight-for-age percentiles

NAME _____

RECORD # _____



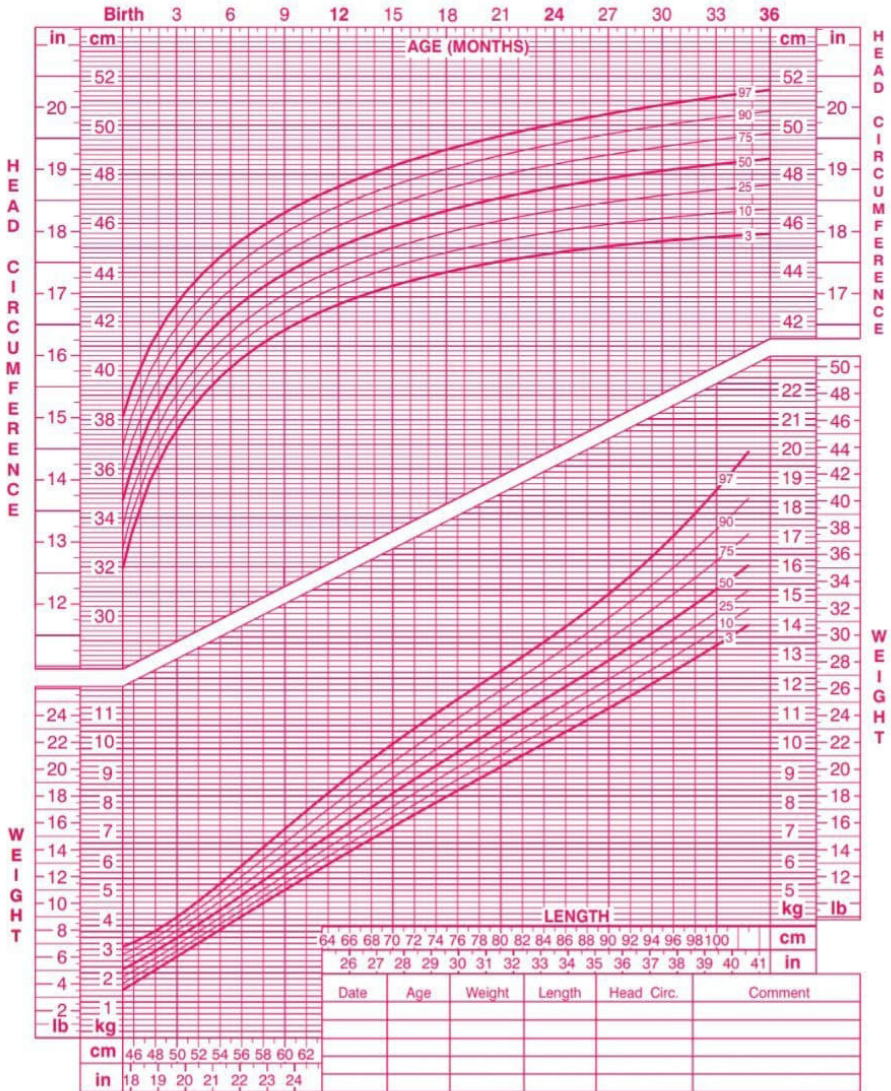
Published May 30, 2000 (modified 4/20/01).
 SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).
<http://www.cdc.gov/growthcharts>



Birth to 36 months: Girls
Head circumference-for-age and
Weight-for-length percentiles

NAME _____

RECORD # _____



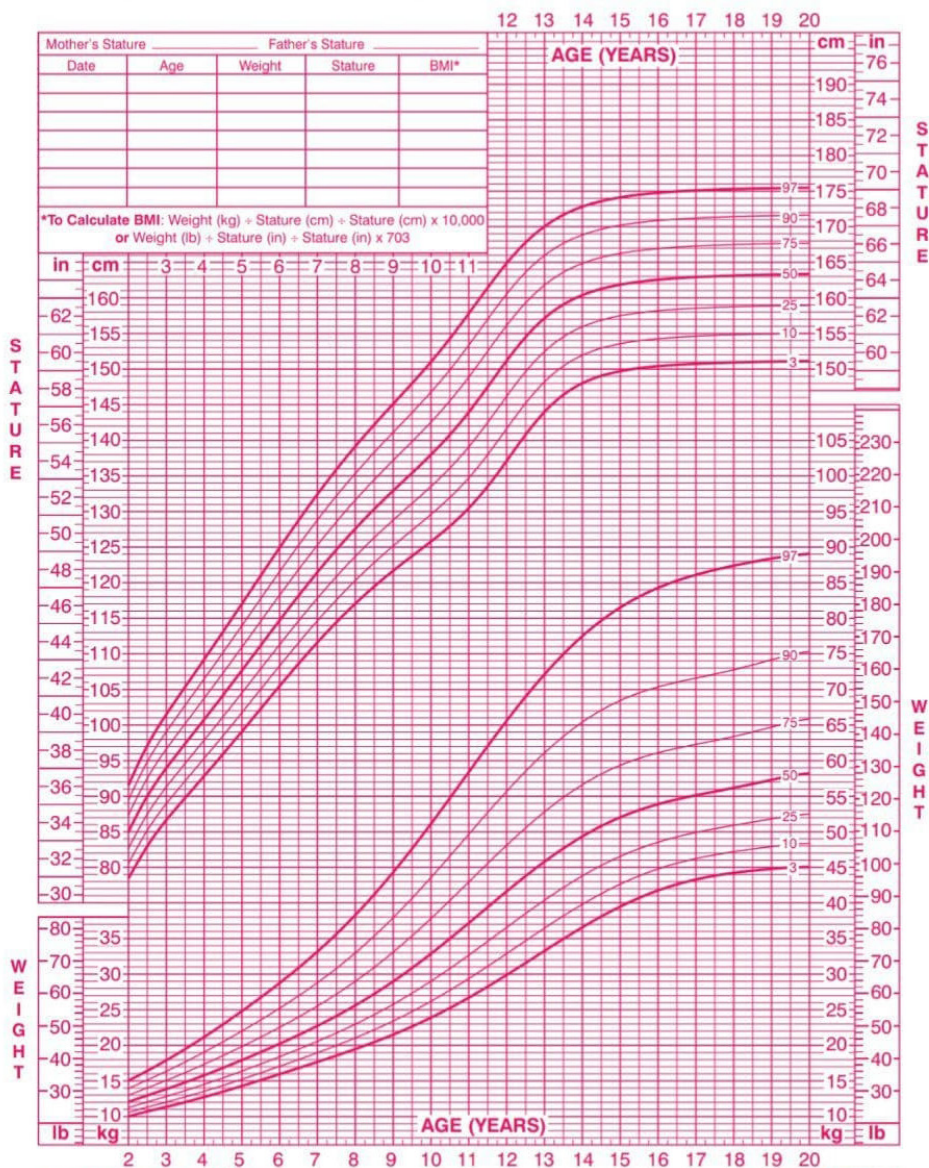
Published May 30, 2000 (modified 10/16/00).
 SOURCE: Developed by the National Center for Health Statistics in collaboration with
 the National Center for Chronic Disease Prevention and Health Promotion (2000).
<http://www.cdc.gov/growthcharts>



2 to 20 years: Girls Stature-for-age and Weight-for-age percentiles

NAME _____

RECORD # _____



Published May 30, 2000 (modified 11/21/00).

SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).
<http://www.cdc.gov/growthcharts>



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Birth history



Past medical history

Admission to ward

No	Date Admitted	Date Discharged	Hospital / Ward	Reason for Admission
1				
2				
3				
4				
5				
6				
7				
8				
9				
10				

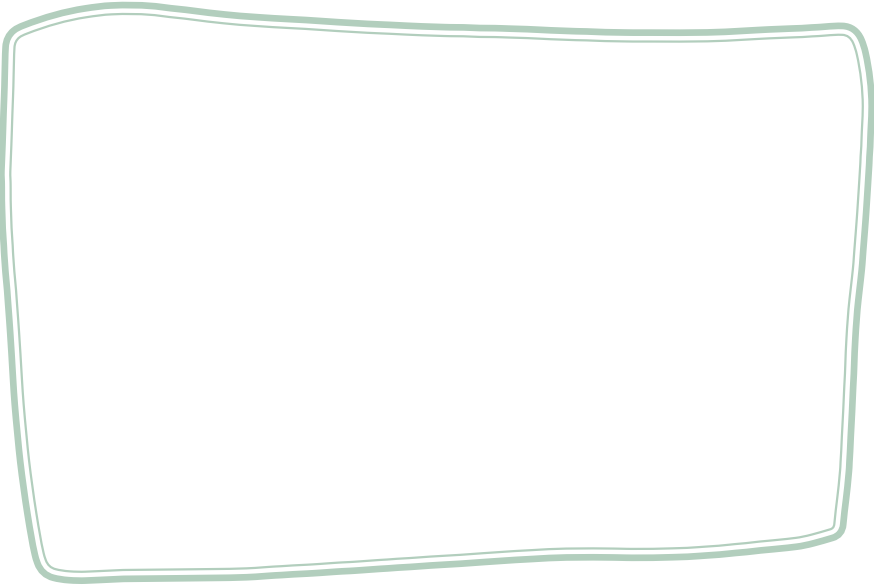
Admission to ward

No	Date Admitted	Date Discharged	Hospital / Ward	Reason for Admission
11				
12				
13				
14				
15				
16				
17				
18				
19				
20				

Past surgical history

No	Date Admitted	Date Discharged	Hospital / Ward	Type of surgery
1				
2				
3				
4				
5				

Family history / Family tree



Social history



Drug history

Current and relevant past medications (drug name, dose, frequency, duration, compliance).

Prescription drugs / Over-the-counter drugs / Traditional medications



Allergy history

Established allergies and allergic reaction / adverse reaction to drugs / food allergies / environmental allergies



Motor function (assessed every six months)

Motor function	Never	Only previously	Currently	Age & Date Assessed	Comments
Holding head up without support					
Rolling onto side					
Sitting without support					
Crawling on hands and knees					
Standing with assistance					
Standing alone (without assistance)					
Walking with assistance					
Walking alone (without assistance)					
Able to walk 10 meters unaided					
Climbing stairs					
Useful function of hands (e.g. hold a pen or pick up a coin)					
Reaching overhead in a sitting position					
Raising hands to mouth in a sitting position					

2.5 Genetic

Common questions parents / caregivers may ask, which can be discussed

- What is SMA? How did my child get it?
- There is no one in my family with SMA. How did my child get SMA?
- What is a carrier? If I am a carrier, can I develop SMA?
- Since SMA is a genetic condition, will my next child be affected with SMA as well?
- Can I prevent my next child from getting SMA?
- What about my unaffected children? How do I know if they are carriers?
- Would a child with SMA be able to attend school?
- Will my child be able to read and write normally?
- Can people with SMA achieve a normal life span?
- What are the major challenges faced by people with SMA?
- What do I do now? Is there treatment available?
- Is there any support groups available?

	Date	Date	Date
Discussion Notes			
Examined by			

2.6 Neurology

	Date	Date	Date	Date
History				
Active complaints				
Current functioning / developmental status				
Feeding / nutrition status				
Current medication (Types, changes in dose, side effects)				
Physical Examination				
General Examination				
Contractures Deformity Posture Muscle bulk				
Others <i>(please specify)</i>				

	Date		Date		Date		Date	
Neurology Examination								
Upper Limb	R	L	R	L	R	L	R	L
Power								
Lower Limb	R	L	R	L	R	L	R	L
Power								

	Date		Date		Date		Date	
Additional Examinations:								
Impression								
Plan								
Examined by								

2.7 Respiratory

Respiratory problem list

- 1.
- 2.
- 3.
- 4.
- 5.

Current respiratory support / equipment

Equipment	Tick (/)	Comments
Bronchodilator		Type: Frequency:
Nebulizer		Frequency:
Cough insufflator- exsufflator		Frequency:
Oral suction		Frequency:
BiPAP		Pressure:

Respiratory review

	Date	Date	Date	Date
History				
Respiratory symptoms (shortness of breath, cough)				
Sleep symptoms (Poor sleep, snoring)				

	Date	Date	Date	Date
Physical Examination				
General Examination				
Work of breathing Paradoxical breathing Cyanosis Cough effectiveness Others <i>(please specify)</i>				
Respiratory Examination				
Inspection • Chest wall deformity Others <i>(please specify)</i>				
Palpation				
Percussion				

	Date	Date	Date	Date
Auscultation				
Investigations				
Chest X-ray				
Sleep study				
Lung function test				
FEV1				
FVC				
TLC				
Overnight pulse oximetry				
Additional examinations				

	Date	Date	Date	Date
Impression				
Plan				
Examined by				

2.8 Rehabilitation & Orthopaedics

	Date	Date	Date
History			
Current physical problem			
Active complaints (pain, weakness, recent fall, wound, rashes, etc)			
Current functional status			
Personal activities of daily living (ADL)			
Instrumental ADL			
Feeding / drinking / lifting objects / wearing own clothes / others:			

Physical Examination			
Deformities			
Neck			
Upper Limb			
Lower Limb			
Muscle Strength			
Upper Limb			
Lower Limb			

2.8 Rehabilitation & Orthopaedics

	Date		Date		Date	
Contractures (ROM, goniometry)	R	L	R	L	R	L
Wrist						
Elbow						
Shoulder						
Hip						
Knee						
Ankle						

	Date		Date		Date	
Spine Examination						
Kyphoscoliosis						
Hyperlordosis						
Hip Examination						
Hip subluxation						
Hip dislocation						
Investigation						
X-ray						
Others						

Additional examination (Motor measures)

	Date	Date	Date
Non-sitters			
Muscle weakness (Antigravity movements)			
Functional scales (CHOP INTEND*)			
Motor development (HINE*)			

	Date	Date	Date
Sitters			
Functional scales: HFMSE*			
RULM*			
MFM*			
(Strength tests)			

	Date	Date	Date
Walkers			
Mobility timed tests			
Muscle weakness (Strength tests)			
Falls Functional scales HFMSE*			
RULM*			

***Abbreviations:**

- Children’s Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND)
- Hammersmith Infant Neurological Examination (HINE)
- Hammersmith Functional Motor Scale Examination (HFMSE)
- Revised Upper Limb Module (RULM)
- Motor Function Measure (MFM)
- 6 Minute Walk Test (6MWT)

Impression			
Plan			
Examined by			

2.9 Nutrition

Assessment	Date	Date	Date
Feeding and Swallowing			
Feeding history <ul style="list-style-type: none"> • Any prolonged feeding difficulty • Meal time, fatigue with oral feeding, evidence of choking or coughing during or after swallowing • History of recurrent pneumonia 			
Mealtime observation			
Examination of oral structures			
Videofluoroscopic swallow studies			

Gastrointestinal Dysfunction			
Symptoms of GERD (emesis, regurgitation, gurgling after feeds)			
History of constipation, abdominal distention and bloating			

	Date	Date	Date
Growth			
Monitor height and weight (Recumbent length, segmental measures, arm span [if unable to stand], contractures)			
Growth velocity			
Body composition (Skin fold measurement, mid upper arm circumference, bioelectric impedance analysis)			

	Date	Date	Date
Other Tests			
Blood work			
Upper gastrointestinal X-ray series			
Impression			
Plan			
Examined by			

2.10 Palliative Care

Palliative care

Topics to cover
Physical condition of patient
Psychological / Mental condition of patient and caregiver
Social well-being for both patient and caregiver
Spiritual well-being for both patient and caregiver
Advanced care plan (RT tube, symptom plan, decision-making would go along the way)
<ol style="list-style-type: none"> 1. Place of care 2. Wishes 3. DNR status 4. End of life planning

	Comments
At diagnosis Date: Reviewed by:	
During crisis Date: Reviewed by:	
At terminal stage Date: Reviewed by:	

Palliative care vision

1. Making choices that are consistent with beliefs and values of parent / caregiver.
2. Choosing what is best for parent / caregiver.
3. Providing support, along with sufficient information, for parent / caregiver.
4. Multidisciplinary approach to provide child and parent / caregiver with the best possible care.
5. Managing aspects of care and ensuring comfort by managing symptoms (e.g. pain, dyspnoea).
6. Managing ethical and legal aspects of care (care planning)

Review of affected body systems, including:

1. Respiratory system
2. Musculoskeletal system
3. Neurology system
4. Gastrointestinal system

Managing mental health of patient and caregiver with assessment of DASS 21 and assessing phase of grief in relation to level of knowledge regarding disease, as well as palliative care.

Evaluating social well-being, including:

1. Patient's kindergarten / school environment.
2. Caregiver's working environment, community, friends and support group.
3. Relationships, such as with siblings and close contacts.

Involving a spiritual approach with respect to the patient's and family's beliefs:

1. Religious ceremony or gathering
2. Beliefs about life and death

ADVANCE CARE DIRECTIVES

This should be formulated through a series of discussions, along with sufficient information given, and set out clearly in a set of documents containing instructions that consent to, or refuse, specified medical treatments that articulate care and lifestyle preferences in anticipating future events or scenarios.

Final objective is to provide the best care, especially at the terminal stage, covering:

- a) Comfort care
- b) Wishes by family / patient
- c) Do Not Resuscitate (DNR) status

Question	Date	Comment
1. Understanding of current condition		
2. Personal or religious belief with regards to resuscitation		
3. Understanding of DNR		
4. Consent and documentation regarding Advanced Care Plan and DNR		
5. Support system and involvement with support group		

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