

**General precautions on respiratory care for COVID-19  
patients with muscular dystrophies, severe physical  
and intellectual disabilities, or neuromuscular  
disorders**

The National Council of Directors of Muscular Dystrophy Facilities

The National Council for Severe Physical and Intellectual Disabilities

The National Hospital Neurology Council

## Preface

When patients with respiratory dysfunction due to neuromuscular disorders such as muscular dystrophy, severe physical and intellectual disabilities, are suspected to have COVID-19, the situation is quite different from that of acute respiratory failure among general COVID-19 patients. In particular, when mechanical ventilation has already been introduced for chronic respiratory failure in patients with these neuromuscular disorders, special attention is required regarding therapeutic considerations and infection control.

We (the National Council of Directors of Muscular Dystrophy Facilities, the National Council for Severe Physical and Intellectual Disabilities, the National Hospital Neurology Council) have developed advice on the treatment of confirmed or suspected COVID-19 patients with neuromuscular disorders requiring respiratory care. This advice was based on the opinions and findings of the facilities participating in the councils. The advice also referred to other international recommendations. We hope that this advice will prove useful in the preparation of protocols at each facility for facing COVID-19 patients.

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## Introduction

To prevent COVID-19, avoiding the so-called 3 Cs of "closed spaces with poor ventilation", "crowded places", and "close-contact settings" appears crucial. Many patients with neuromuscular disorders (NMDs) such as muscular dystrophies (MDs) and severe physical and intellectual disabilities (SPID), however, need assistance in their daily lives and close contact is thus unavoidable with these patients. When these patients require medical care, the number and types of caregivers involved will differ depending on each situation. Among the care and treatment required, respiratory care such as mechanical ventilation (MV) and cough augmentation is essential to maintain patient health and quality of life (QOL). On the other hand, such treatments are known to scatter aerosols<sup>1-4</sup>. Dealing with a patient suffering from COVID-19 (including suspected patients) raises concerns about the risk of infection not only to the caregiver and/or medical staff, but also to other people in contact with these personnel. Especially in nursing homes and medical care wards, which care for many high-risk patients, prevention of nosocomial infection is paramount. This document describes the risk of droplets and aerosols in respiratory care for patients with NMD, and outlines general countermeasures for nursing homes and medical care wards, etc., with reference to studies and recommendations that have already been issued<sup>1,3</sup>.

### Risk of droplets and aerosols during respiratory care of patients with NMD

- 1) In patients with NMD, respiratory dysfunction causes type II respiratory failure (alveolar hypoventilation) and reduces airway clearance.

- 2) Currently, non-invasive ventilation (NIV) is mainly used for type II respiratory failure. However, NIV involves a non-closed circuit, and thus cannot avoid aerosol diffusion<sup>1,3,4)</sup>.
- 3) Even in MV with a tracheotomy (tracheal intermittent positive-pressure ventilation; TIV), a closed circuit cannot be maintained in patients with severe tracheal deformity, especially when a speech cannula or loosely adjusted cuff has been used. As a result, leakage will occur and aerosols will be dispersed around the patient.
- 4) Respiratory physiotherapy such as lung inflation and cough augmentation, mechanically assisted cough (MAC), and suction are extremely important for the prevention of secondary lung injury, but dispersal of droplets and aerosols with these treatments cannot be avoided.
- 5) These procedures are performed by the caregiver in close proximity to the patient, so when the patient has developed COVID-19, the caregiver will be exposed to massive doses of infected droplets/aerosols. Therefore, in respiratory care for confirmed or suspected COVID-19 patients, we should recognize the risks of droplets and aerosols, and should reduce these risks using appropriate precautions.

### Leak points in NIV and countermeasures against them

- 1) In NIV, leaks can occur from the site of mask fitting, the mouth of nasal mask users, or the exhalation port. When the leak is large, inspiratory air flow from the ventilator could be increased to maintain the set ventilation. A large amount of aerosol is then likely to be dispersed, and the environmental pollution becomes serious<sup>1)</sup>. As a countermeasure, in confirmed or suspected COVID-19 patients, use of a full-face mask (or total face mask) that covers both the nose and mouth and fits the shape of

the face is highly recommended (Fig. 1a,b)<sup>1-3</sup>). A helmet-type mask that covers the entire head is also available (Fig. 1c), but positive pressure is difficult to maintain with the air supply capacity of most of the currently used portable types of ventilator, so a ventilator running off compressed air would be preferable. As ventilation conditions will differ when the mask or circuit is changed, it is necessary to check the breathing condition and adjust ventilator settings accordingly.

Fig. 1. Examples of full-face, total face, and helmet-type masks

a.



b.



c.



a, b. Non-vented disposable masks (Classic Star® Dräger)

Full-face mask® (a. left), Total face mask® (a. right)

Suppression of leaks is easily achieved by adjusting the inflation of the air cushion providing the seal (b)

c. Helmet-type mask (StarMed CaStar®; Intersurgical)

Referring to the homepage of Intersurgical

(<https://www.intersurgical.com/products/critical-care/starmed-respiratory-hoods-for-non-invasive-mechanical-ventilation-niv>)(2020/6/16)

- 2) In NIV, a single circuit with an expiratory port is widely used. In such cases, aerosols are constantly scattered from the expiratory port. When a vented mask is used, the exhalate is disseminated into the room air, so a non-vented mask should be used, placing a filter with excellent virus-removal performance (hereinafter, “V filter”) between the mask and exhalation port (Fig. 2a). This will reduce the amount of virus diffusion. When a vented mask must be used, it may be possible to cover the port with a non-woven mask, but care should be taken to avoid blocking exhalation itself.
- 3) In a single circuit with an exhalation valve, place a V filter between the mask and exhalation valve (Fig. 2b). Avoid using a heating humidifier when using the V filter.



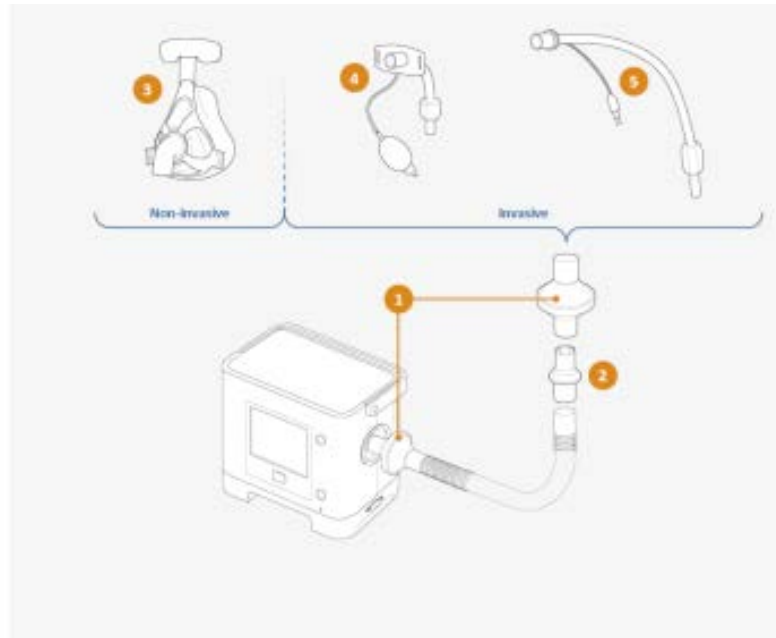
Fig. 2. Examples of single circuits

a. A single circuit with an expiratory port

Passive circuit:  
Option 1

- 1 Bacteria/Viral filter  
With this option, if limited supply of filters, the one at the device outlet could be omitted as long as the filter between the interface and leak port remains.
- 2 Exhalation leak port
- 3 Non-vented (without integrated leak) NIV mask
- 4 Tracheostomy tube  
(trach adapter not shown and optional to connect to circuit)
- 5 Endotracheal Tube (ETT)  
(trach adapter not shown and optional to connect to circuit)

Having extra filters or an HME in the circuit may affect performance of alarms and an increase in pressure drop at the patient may be experienced.

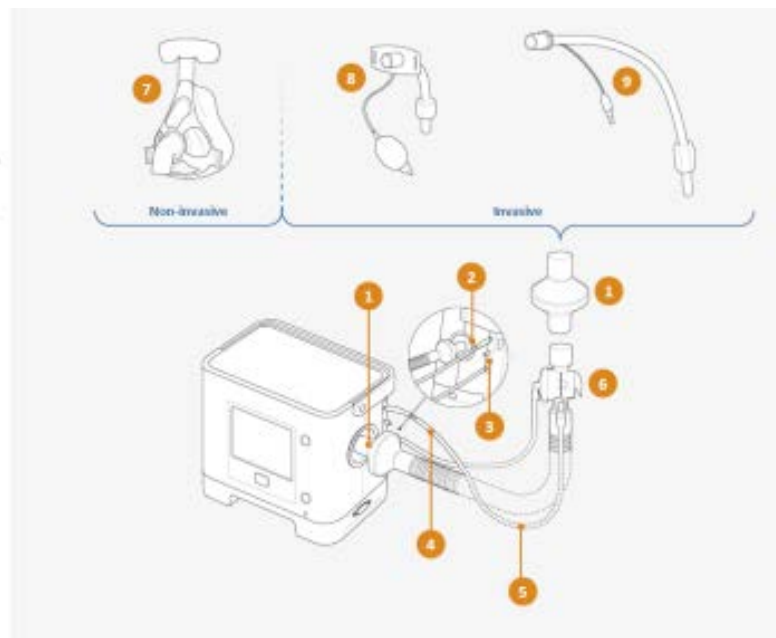


b. A single circuit with exhalation valve

Active PAP circuit:  
Option 1b

- 1 Bacteria/Viral filter  
With this option, if limited supply of filters, the one at the device outlet could be omitted as long as the filter between the interface and exhalation valve remains.
- 2 Trilogy universal active PAP tube adaptor
- 3 Universal porting block (3 ports)
- 4 Proximal pressure line
- 5 Active exhalation valve line
- 6 ActivePAP circuit
- 7 Non-vented (without integrated leak) NIV mask
- 8 Tracheostomy tube  
(trach adapter not shown and optional to connect to circuit)
- 9 Endotracheal Tube (ETT)  
(trach adapter not shown and optional to connect to circuit)

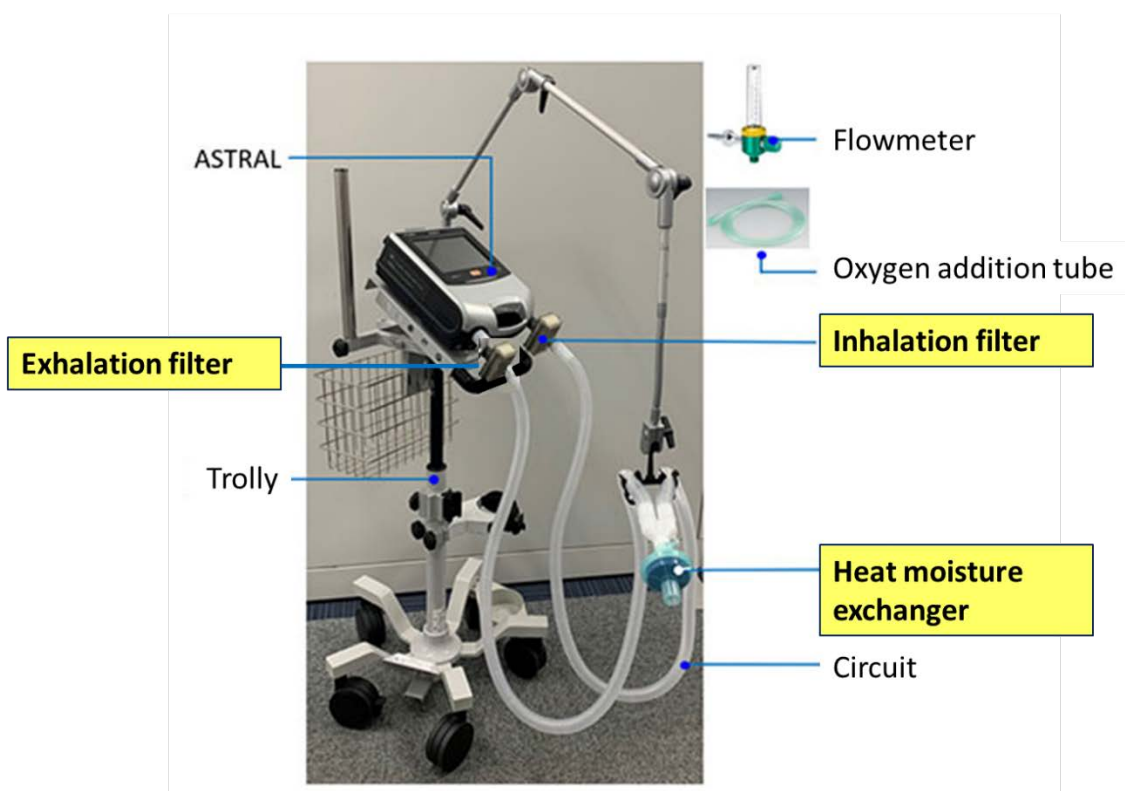
Having extra filters or an HME in the circuit may affect performance of alarms and an increase in pressure drop at the patient may be experienced.



Referring to Trilogy platform COVID-19 example circuit configurations (Philips)

4) Where possible, it is advisable to use a double-branch circuit (Fig. 3) with V filters on both inspiratory and expiratory sides<sup>1,3-5</sup>). A double-branch circuit is a standard configuration in critical areas such as intensive care units, and is therefore recommended for NMD patients as a preventive measure against the infection, although the number of portable ventilators available at present is limited<sup>4,5</sup>).

Fig. 3. An example of a double-branch circuit (CleanAir ASTRAL™ 150® ResMed)



Referring to the homepage of Fukuda Denshi Co Ltd. (partly modified by the authors)

([https://www.fukuda.co.jp/medical/products/inhome\\_medical/astral.html](https://www.fukuda.co.jp/medical/products/inhome_medical/astral.html))

(2020/6/16)

5) Use of a heat moisture exchanger (HME) with bacterial filter function is appropriate for humidification. When using a heating humidifier, use the automatic water supply module to prevent the risk of aerosol diffusion during water supply<sup>4)</sup>.

6) Additional installation of the V filter and HME increases dead space and circuit resistance, which may alter ventilation conditions, evoke failure to detect spontaneous breathing, and result in malfunction of the out-of-circuit alarm. Circuit resistance may also rise further due to contamination and water retention, as usage time increases. For these reasons, vital monitoring such as percutaneous O<sub>2</sub> saturation (SpO<sub>2</sub>) as well as respiratory alarms is recommended in the implementation of these procedures<sup>3,4)</sup>.

7) Most types of portable ventilators take room air, so when a patient is suffering from COVID-19 and aerosols containing the virus are dispersed, the inside of the ventilator will be contaminated. If available, selecting a ventilator that takes compressed air from the central piping is ideal. Use of a ventilator model with an air intake filter that has the ability to remove viruses is also preferable<sup>4)</sup>, but the number of such models in distribution is limited. When these measures are not available, the ventilator used for a confirmed or suspected COVID-19 patient must be solely dedicated to that individual patient<sup>4)</sup>. The circuit with ventilator must be disposable and should be replaced according to an appropriate procedure to minimize aerosol diffusion<sup>4)</sup>.

#### Leak points in TIV and countermeasure against them

1) In TIV, a closed circuit has basically been formed, but many NMD patients use loosely adjusted cough or speech cannulae to maintain vocal communication, and leakage will thus occur from the mouth, nose and parts of the tracheostomy.

2) Open-type suction involves a further problem, in that aerosols are scattered during the procedure. As a countermeasure, the respiratory circuit should be arranged in the same way as NIV. Properly manage the cuff pressure to maintain a closed circuit<sup>6)</sup>. Use a closed tracheal suction system to prevent diffusion of aerosols during suction, or use a double suction tube to reduce the frequency of suction.

### Precautions for oxygen therapy

1) Oxygen monotherapy without MV should be avoided in patients with NMD. When required, it is preferable to use a face mask rather than a nasal cannula and to cover it with a surgical mask, in order to prevent aerosol diffusion<sup>3)</sup>, but the risk of CO<sub>2</sub> retention becomes markedly higher.

2) When oxygen is used, monitor the ventilation status by transcutaneous CO<sub>2</sub> monitoring or regular blood gas tests, and when any concern about CO<sub>2</sub> narcosis arises, immediately consider introducing MV.

### Precautions for MAC

1) Currently, several models are used as sputum-assisting devices. Their main functions are mechanical insufflation-exsufflation (MI-E), intrapulmonary percussive ventilation (IPV), and high-frequency chest wall oscillation (HFCWO). MAC is extremely important for maintaining airway clearance in NMD patients, but careful judgement of the necessity and urgency is warranted for confirmed or suspected COVID-19 patients and the maximum protective measures should be taken, since MAC produces and disseminates large amounts of aerosols.

2) MI-E induces coughing by applying positive pressure to the airways for a few seconds, then shifting to negative pressure to promote sputum production. In cases using a mask, when adhesion is insufficient, leakage may occur from the mounting portion in the positive phase. Even in cases of tracheotomy, leakage may occur if cuff pressure is insufficient. Further, contamination of the inside of the device becomes a problem, since the exhalate of the patient is sucked into the device during the negative phase. Keep the mask suitably tight and properly adjust the cuff pressure to minimize leaks and place a V filter between the mask/tracheal cannula and circuit.

3) For IPV, place a V filter between the expiratory port and mask/tracheal cannula and port.

4) In HFCWO, cover the percussion wrap attached to the chest wall with vinyl so that it does not come into direct contact with the patient<sup>5)</sup>. Covering the patient's head with a vinyl tent, etc., is expected to suppress aerosol diffusion to some extent, but evidence is currently lacking. The sputum support device has a risk of contaminating the inside of the device, because it takes room air regardless of the mode. Thus, limiting its use to individual COVID-19 patients is necessary.

### Precautions for respiratory physiotherapy

1) For patients with NMD, respiratory physiotherapy such as lung inflation with resuscitation bags and cough augmentation are performed. Air leaks from masks and droplets/aerosols dissemination associated with cough also occur during these treatments, so the necessity of physiotherapy is carefully judged in confirmed or suspected COVID-19 patients. When using a resuscitation bag, attach a V filter between the mask/cannula

and the resuscitation bag and exclusively use the resuscitation bag for the COVID-19 patients<sup>4)</sup>.

2) Protective measures should be taken during lung inflation and cough augmentation, such as having the patient wear a mask and directing the face to the side opposite the caregiver during exhalation, if possible. Adequate ventilation is important during and after respiratory physiotherapy.

3) In NMD patients, respiratory physiotherapy is extremely important, even when suffering from COVID-19, but treatment should be minimized in consideration of the infection state and sputum status, and alternative treatments such as postural drainage should be carefully considered<sup>3,7,8)</sup>.

## Consideration of the environment surrounding patient rooms and methods of preventing caregiver infection

1) Active ventilation, isolation of confirmed or suspected COVID-19 patients from non-infected patients, appropriate use of personal protective equipment (PPE), and environmental cleaning should be carefully considered.

2) The first step is to perform clear zoning. The rooms of COVID-19 patients must be designated as infected areas, and a semi-clean area should be placed between infected areas and clean areas. Clearly divide each area with a tape or partition to prevent viruses from being brought into the clean area. This is particularly important when COVID-19 and non-infected patients are together within the same ward or facility, and care should be taken not to cross the infected and clean areas.

3) For air ventilation, isolation in a negative-pressure compartment is ideal, in consideration of the risk of constant aerosol diffusion in patients with NIV or tracheostomy with a semi-closed circuit. Even if this is difficult, the confirmed or suspected COVID-19 patient should be isolated in a private room as much as possible, and adequate air ventilation should always be performed. This entails a certain unavoidable cost, but it is preferable to create a negative-pressure space using a simple negative-pressure device. If air flow from the infected area to semi-clean or clean areas is difficult to control, consider blocking the air flow with a vinyl curtain.

4) Caregivers should wear N95 masks when entering infected areas. Wear PPE such as gloves, face guard, vinyl apron (arm cover) and cap when performing care<sup>1,4,9)</sup>. To prevent contact infection, thoroughly clean the hands and perform hand sterilization at the time of entry/exit and care, regularly perform environmental cleaning (disinfection) of the patient rooms and infected areas. Items in the infected area should not be taken out, in principle. When such items must be taken out, thoroughly disinfect them. Time and effort is required to remove PPE, and the caregiver is likely to be infected when undressing. It is important that two members form a pair and check each other to ensure appropriate removal.

5) Consider using telecommunications such as intercoms and videophones to avoid unnecessary entry and stay in the infected room. In addition, individual caregivers should avoid caring for both COVID-19 and non-infected patients within the same shift.

Things to be aware of when implementing infection control

1) To date, we have described general measures, but actual implementation of these measures involves many difficulties and issues to be considered.

2) The first is the serious lack of supplies caused by the global pandemic of COVID-19. In a questionnaire survey of 12 facilities in the Japanese National Hospital Organization, which has special wards for patients with MD and SPID, serious concerns were raised regarding the supply of ventilators, tracheal cannulae, ventilator circuits, HMEs and HEPA filters. As a result, most facilities have taken or have been considering measures such as postponing the replacement cycle or using alternatives (Table 1). A serious supply shortage of PPE was also seen. N95 masks are indispensable in the care of patients at high aerosol risk<sup>1,4,9)</sup>, but securing a stable supply has been difficult.

Table 1. Impact of COVID on supply of ventilators and related products (Urgent investigation of 12 facilities related to supply of ventilators and related products during pandemic condition of COVID-19)

Facility	Ventilator	Circuit, HME	Tracheal cannula	Enteral tube
1	○	×	×	
2		△	○	○
3		×	×	○
4	○	△	×	×
5		×	×	×
6	○	△	△	



7	○	×	○	○
8	△	×	×	
9		×	○	○
10		×	○	
11	×	×	×	×
12	×	×	×	○

○, As usual; △, considering measures (postponing the replacement cycle or using alternatives, etc.); ×, lack of supplies or taking measures; Blank, no answer.

3) The second is a structural problem with facilities. Most hospitals and institutions that treat patients with MD and SPID do not have a negative-pressure room, etc. Furthermore, many facilities are obliged to treat COVID-19 and non-infected patients within the same ward, and proper zoning seems difficult in some facilities.

4) In addition, many problems are seen on the side of patients and also in terms of ethical issues. In NMD patients, where respiratory care is essential, daily treatment and care should be continued even under pandemic conditions of COVID-19<sup>2,6,10)</sup>. In the case of confirmed or suspected COVID-19 patients, a comprehensive judgment will be made considering the necessity of treatment, the medical condition and the environment. Some patients have developmental disorders, mental retardation, or dementia, and many patients thus cannot accept changes even if it is necessary to change the ventilator system or device in terms of infection control and treatment of infectious diseases. Concerns have also been raised regarding mental stress associated with isolation and restrictions on visits by family members<sup>6)</sup>.

5) Infection control always involves an ethical dilemma, because of the constraints placed on individual freedom and QOL. In particular, how much treatment is appropriate when a positive contact is suspected due to onset in a family member or caregiver is an annoying problem. How best to adjust the many contradictory issues such as the life and QOL of the patient, the safety of caregivers and other patients, and the maintenance of medical functions may differ depending on the situation, the condition of the patient, the environment, etc. In actual application, it is important to take the best possible measures according to individual circumstances, taking into consideration the infection status, the condition of the general population, the necessity of respiratory care, the environment, and collective immunity status.

6) On the other hand, COVID-19 is likely to become serious in NMD patients with reduced respiratory function and swallowing dysfunction<sup>2,10)</sup>. With COVID-19, the respiratory status may rapidly deteriorate even in the general population in a short time, so changes in respiratory management methods and endotracheal intubation are likely to be necessary. Endotracheal intubation is often difficult in patients with MD/SPID due to spinal/thoracic deformity and difficulty opening the mouth, so emergency intubation should be avoided. If limits to NIV support exist, intubation should be performed during a time when sufficient human resources can be secured<sup>3)</sup>.

7) Consideration should be given to the use of a video laryngoscope, as intubation procedures carry the risk of aerosol dissemination. To prevent droplets/aerosols from directly hitting the operator, consider a device such as covering the head with an aerosol box (Fig. 4).

Fig. 4. Example of an aerosol box



Referring to the blog of the intensive care unit at Kameda Medical Center

([http://www.kameda.com/pr/intensive\\_care\\_medicine/post\\_59.html](http://www.kameda.com/pr/intensive_care_medicine/post_59.html)) (2020/5/18)

### Closing remarks

Clusters of COVID-19 have been formed in medical hospitals/institutions and nursing homes. In medical wards and nursing facilities that treat many NMD patients requiring respiratory care, precautions against infection within the facility are extremely important. We hope you find this helpful in taking appropriate precautions while maintaining daily care.

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