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Q & A with Dr. Priya Kishnani (Duke University*) on COVID-19 and Pompe Disease

*The following advice is based on the information available as of **3/19/2020**. The COVID-19 pandemic is a rapidly evolving situation; please check WHO, CDC, and the State Health Department websites routinely for the latest updates, guidelines, and recommendations.*

The information below is for informational purposes only. The AMDA and Dr. Kishnani and her Pompe team strongly advise that you discuss all information with your physician.

General Questions:

1) Are Pompe patients considered “high risk” according to the CDC (United State’s Centers for Disease Control and Prevention) and WHO (World Health Organization) guidelines?

In general, we would consider patients with Pompe disease to be at high risk due to their chronic medical condition.

Factors that would further increase the risk of infection and/or progression of COVID include:

- a) Any form of secondary infections or conditions considered high risk by CDC
- b) Currently or recently on immune modulation, without evidence of B-cell recovery (Please refer to Question# 2)
- c) Recent hospitalizations
- d) Insufficient respiratory and/or cardiac function

Other factors that may increase this risk in individuals with late-onset Pompe disease (LOPD) are:

- a) Age \geq 65 years
- b) Upright FVC $<$ 55% predicted
- c) A significant drop in supine FVC (or supine FVC $<$ 25% predicted),
- d) Ventilator dependence or use of assistive devices for breathing such as BiPAP, CPAP, etc.)
- e) Other general risk factors: Hypertension, diabetes, asthma, COPD (please refer to CDC)
- f) Pregnancy

Other factors that may increase risk in individuals with infantile Pompe disease (IPD) include:

- a) Significant hypertrophic cardiomyopathy and other cardiac complications such as heart failure
- b) New diagnosis of Pompe disease
- c) Failure to thrive
- d) History of aspiration
- e) Ventilator dependence (including use of CPAP, BiPAP, etc.)

2) Is my immune system compromised because of Pompe or is it like every other person?

Although Pompe disease does not directly impact the immune system, special considerations should be taken in individuals who have received immune-modulatory drugs such as rituximab, methotrexate, bortezomib, sirolimus, rapamycin and others. Individuals who are either currently receiving such treatment or have received in the past year should consult with local treating physician regarding lymphocyte and immunoglobulin levels. Individuals with a normal lymphocyte count, i.e., who have a B-cell recovery would not be considered immunocompromised.

3) Am I “high risk” only because of decreased respiratory function? If I have Pompe and normal respiratory function, am I still high risk?

Decreased respiratory function is not the only reason for an individual to be considered ‘high risk’. There are a number of other factors that can put an individual in the ‘high risk’ category, including immune modulation, reduced pulmonary and cardiac function, etc. (Please refer to Question# 1 and CDC guidelines).

4) What precautions beyond the CDC and WHO Guidelines do you recommend Pompe patients take?

Apart from the CDC and WHO guidelines,

- (a) Keep yourself informed about the latest development on coronavirus in your local community
- (b) Family members: Family and friends who are in close contact should also be taking all the necessary precautions to avoid transmitting COVID-19 to an individual with Pompe disease.
- (c) Avoid falling prey to misinformation and deceptive products. Check with your doctor before starting any new medications to ‘prevent’ COVID-19. Check on WHO webpage on the misconception about spread and treatment of coronavirus-<https://www.who.int/emergencies/diseases/novel-coronavirus-2019/advice-for-public/myth-busters>. FDA has a website listing fraudulent COVID-19 products (<https://www.fda.gov/consumers/health-fraud-scams/fraudulent-coronavirus-disease-2019-covid-19-products>).
- (d) Individuals who are currently receiving immune modulation or have recently completed immune modulation with lack of recovery of the immune system are at a higher risk, and should consider self-isolation as much as possible (Please refer to Question# 2).
- (e) Facemask: If you are healthy, you only need to wear a mask if you are coughing, sneezing or feeling unwell, or taking care of a person with suspected COVID-19 infection. Furthermore, masks are effective only when used in combination with frequent hand-cleaning with alcohol-based hand rub or soap and water. If you wear a mask, then you must know how to use it and dispose of it (<https://www.who.int/emergencies/diseases/novel-coronavirus-2019/advice-for-public/when-and-how-to-use-masks>)

5) Is it safe to go to grocery store if I need to? Alternatively, if I have my groceries delivered, do I need to wash the cans?

It should be okay to go to the grocery store if you do not have other risk factors (outlined in Question#1). However, practice social distancing (6 feet or 1-2 meters) and self-isolation as much as possible to avoid exposure to coronavirus. Getting your groceries and other

essentials via either a delivery service or curbside pick-up will be a safer alternative. As per the WHO guidelines, the risk of coronavirus infection transmission from the surface of a commercial good is low, so washing/wiping surfaces may not be required (more details available in FAQs: <https://www.who.int/news-room/q-a-detail/q-a-coronaviruses>).

6) Will wearing a facemask protect me from contracting the virus?

Based on the CDC recommendations, we do not recommend wearing masks to prevent COVID-19. If you have an acute respiratory illness and have symptoms such as cough or fever, you should wear a mask. It is VERY important that you follow proper precautions with wearing and disposing of the mask. The WHO has excellent information about how to properly wear and dispose of a mask. Please consider watching the following videos to properly wear a mask: <https://www.who.int/emergencies/diseases/novel-coronavirus-2019/advice-for-public/when-and-how-to-use-masks>

7) If I have a scheduled appointment with my doctor for routine follow-up or to re-authorize my infusions for another 6 months, what should I do?

You should talk with your local physician to discuss the precautions they are taking at their office to minimize risks to patients. The ideal solution would be if tele-medicine appointments could be arranged.

Infusion Related Questions:

1) Should patients consider skipping infusions to maintain full isolation at home?

We do not recommend missing infusions. For those receiving home infusions, please ensure whoever enters your home is not ill and has limited access within your home. Social distancing should be maintained when appropriate. If you or someone in your household is ill, you should inform your infusion nurse. If you are receiving infusions at a hospital or outpatient infusion center, you should continue infusion treatments at that location and determine if the infusion can be changed to a home infusion setting. If you are ill, you should consider rescheduling your infusion. It is not recommended to receive enzyme infusions while you have an acute illness including fever. If you are receiving ERT under a clinical trial, please keep in close contact with your study site about continuing treatment under the study and options available, such as moving to home infusions, to ensure infusions will not be missed.

2) Are there special precautions (outside of the ones mentioned above) that I should take during an infusion?

None other than the precautions recommended by the CDC and WHO, listed above.

Pompe and COVID-19:

1) If I believe that I have been exposed to COVID-19, what should I do?

- (a) If you have symptoms or have been exposed to COVID-19, you should notify your primary healthcare provider by phone. Also, there may be telehealth options in your area / for your health system.
- (b) Remember to call your doctor first, before going to a clinic, Emergency Department, or Urgent care center. This will enable the clinic to make sure you are isolated to protect other patients.
- (c) If you have asthma, it is important that you use your controller medication every day, even if you have no symptoms, and have your rescue inhaler on hand at all times.

- (d) Please follow CDC guidelines for ‘if you are sick’ - <https://www.cdc.gov/coronavirus/2019-ncov/if-you-are-sick/steps-when-sick.html>. The website also has a Coronavirus self-checker where you can put in your symptoms and it may be a good guide to direct you towards next steps, if your healthcare provider is not available immediately.
- (e) Please call 911 if you have a medical emergency and inform the dispatch if you have symptoms of Covid-19.

2) If I think I have been exposed, how do I explain that I am high risk when I don't look sick?

Given you have Pompe disease with underlying pulmonary compromise, explain to your treating physicians that your response to and progression of COVID-19 illness may be poorer than other individuals without underlying comorbidities. However, the treatment for COVID-19 infection would not differ for individuals with Pompe disease. If available, keep your medical file with you where your diagnoses and medications are listed. This can often be obtained via a MyChart website.

3) I have heard that the pneumonia vaccine will work against COVID-19. Is this true?

Vaccines against pneumonia and flu, such as pneumococcal vaccine and influenza type B, do not provide protection against the COVID-19. There is on-going research to develop a specific vaccine against the COVID-19.

Although the pneumococcal and flu vaccines do not protect against COVID-19, these vaccines are beneficial in general and provide protection from other respiratory illnesses, especially in individuals with Pompe disease.

4) If I am diagnosed with COVID-19, is there anything specific I should tell the treating doctors?

Inform your treating doctor about your Pompe disease diagnosis as well as any other respiratory and heart diseases or other medical issues that you may have. One may have to explain certain details of Pompe disease; where individuals are known to have problems in their diaphragm causing difficulty in breathing when supine (lying down), an increased susceptibility to pulmonary complications, and a possibility of delayed or worsened response to routine treatments for COVID-19 based on the Pompe disease.

For IPD and LOPD, there are increased risks of aspiration, tongue weakness, progression from upper respiratory infection to lower respiratory infection (URI to LRI), anesthesia risks, and difficulty in coming off ventilators. The following website provides the current guidelines for treating an individual with Pompe disease, emphasizing on all of these aspects. Consider keeping this information available, if you visit a doctor for COVID-19 management (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3110959/>)

5) If I get COVID-19, will it kill me?

It is difficult to predict outcomes in any individual at this time. Outcomes depend on various factors such as baseline overall health status (see CDC for details of individuals considered to be at a higher risk), baseline disease status, how an individual reacts to the treatments, other complications, availability of ventilators, etc. Take all the recommended precautions if you live in close proximity with others, and ensure they are also taking necessary precautions because you would be considered a ‘high risk’ individual in the current situation (Refer to Question#1).

At present, the mortality rates with COVID-19 have been linked to age. Individuals over 80 years of age have the highest risk of fatality (about 18% do not do well). Stay updated with the CDC and WHO guidelines to prevent infection and complications (<https://www.cdc.gov/coronavirus/2019-ncov/index.html>).

General advice:

- 1) Wash your hands often with soap and water for at least 20 seconds, especially after going to the bathroom; before eating; after blowing your nose, coughing, or sneezing; and after coming home from a walk or groceries.
 - a) If soap and water are not readily available, use an alcohol-based hand sanitizer with at least 60% alcohol.
 - b) Everyday habits that can help prevent the spread of several viruses can be found on the CDC website <https://wwwnc.cdc.gov/travel/notices/warning/novel-coronavirus-china>.
 - 2) Clean surfaces frequently (don't forget door handles)
 - 3) Latest reports suggest that people with mild symptoms may be contagious. Therefore, social distancing (as recommended by the CDC and WHO) should be followed by people with Pompe and the family they are in close contact with
 - 4) Continue routine exercise (Refrain from starting a new regimen or program)
 - 5) Avoid any elective medical/surgical procedure
 - 6) Stay well hydrated
 - 7) Ensure that all your medications at home are in date (not expired for use). Have 2-3 months of prescriptions on hand if possible
 - 8) Maintain a copy of your written emergency guidelines and diagnoses of Pompe disease and other comorbid illnesses handy to show to other healthcare provider. You could also keep a copy of this Q and A available.
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Note:

*The views and opinions in this document are those of the authors only. They do not convey the views and opinions of Duke University.