World PI Week Quiz

Answers

Here is the 'cheat sheet' to assess your understanding of primary immunodeficiency!

1. How many people are estimated to live with primary immunodeficiency worldwide?

Answer: b.
It is estimated that 6 million people worldwide are living with primary immunodeficiency.

2. Is primary immunodeficiency an infectious disease?

Answer: b.
There are two main types of immunodeficiency: primary immunodeficiency, and secondary (acquired) immunodeficiency. Primary immunodeficiencies are rare diseases which occur when a person's immune system is absent or does not function properly. They are inherited defects of the immune system (carried through the genes). Secondary immunodeficiency is the result of disease or environmental factors such as HIV, malnutrition, or medical treatment weakening the immune system. Some types of secondary immunodeficiency are infectious.

3. How many different types of primary immunodeficiency exist?

Answer: d.
There are over 320 forms of primary immunodeficiency (PI or PID), ranging widely in severity.

4. How many people living with primary immunodeficiency are undiagnosed worldwide?

Answer: c.
It is estimated that 6 million people worldwide are living with primary immunodeficiency. 70-90% of people with primary immunodeficiencies are still undiagnosed worldwide.

5. Which of the following are potential signs and symptoms of primary immunodeficiency? (more than one is correct)

Answer: a, b & c.
A number of different signs and symptoms can help detect primary immunodeficiencies as they occur commonly in many types. The most common are severe, persistent, unusual and recurring infections such as ear, sinus, or skin infections, as well as inflammation in the lungs, liver and intestines. Although it varies from individual to individual and from disorder to disorder, those signs can enable doctors to refer the patient for further medical examinations. It is also important that family carers and the public are aware of symptoms to visit an immunologist in case of suspicion.

6. Are the warning signs of primary immunodeficiency defined?

Answer: b.
Primary immunodeficiency can be strongly suspected in individuals with a family history. Beyond, the warning signs have been characterised as a history of infections which are Severe (requiring intravenous antibiotics and hospital admission), Persistent (difficult to treat with standard regimens), caused by Unusual infective organisms (opportunistic pathogens) and Recurrent (repeated infection at the same site or with the same organism).

7. What are the main reasons for a delayed or misdiagnosis of primary immunodeficiency? (more than one is correct)

Answer: All.
Primary immunodeficiency often presents itself in the shape of “common” infections, sometimes leading physicians to treat the infections while missing the underlying cause. This allows the infections to reoccur, and leaves the patient vulnerable to vital organ damage, physical disability, and even death.

8. Treatment options for primary immunodeficiency: which of the following are possible treatments?

Answer: All.
All can be potential treatment options for a patient who require a very individualised approach.

9. What are immunoglobulin therapies?

Answer: All.
Immunoglobulins are made from human plasma and replaces the protective antibodies that are missing in the immune system, without which patients are prone to recurring and severe infections. They can be administered either intravenously or subcutaneously.
As plasma derived medicinal products, plasma and blood donations are crucial to produce immunoglobulin therapies. Patients rely on the commitment of plasma donors.

10. Is it possible to screen newborns for primary immunodeficiency?

Answer: b.
For severe forms of primary immunodeficiency, newborn screening is a lifesaving, cost-effective method to ensure early diagnosis and save lives. Routine newborn screening programmes for severe primary immunodeficiency (e.g. SCID and XLA) should be widely implemented in public healthcare settings in all countries.

11. When was primary immunodeficiency first classified?

Answer: b.
Primary immunodeficiencies were initially classified in 1970 by a committee of the World Health Organization. At the time, 16 types were identified. By 1998, the number had reached 50. In 2018, more than 320 have been categorized.