



Australian Government

Department of Health

Acting Chief Medical Officer

**FOR INFORMATION: IFNAR1 DEFICIENCY AND SERIOUS ADVERSE EVENTS
FOLLOWING IMMUNISATION**

Essential information:

IFNAR1 deficiency is a rare inherited condition affecting some people in Australia of Western Polynesian heritage including Tongan, Samoan, and Niuean people.

It is associated with severe illness and death from certain viral infections and also potentially from live-attenuated virus vaccines, including the measles, mumps, and rubella (MMR) vaccine.

Currently, the diagnosis of IFNAR1 deficiency prior to vaccination is difficult unless a person has been diagnosed following a severe illness secondary to a viral infection in early childhood. There is no screening genetic test currently available for IFNAR1 deficiency.

The Australian Technical Advisory Group on Immunisation (ATAGI) has prepared a statement on IFNAR1 deficiency. The statement does not recommend any changes to the National Immunisation Program.

ATAGI recommends that all people in Australia, including people of Tongan, Samoan, and Niuean heritage, continue to receive the MMR vaccine given that illness from measles and mumps infections is more severe in unvaccinated individuals, including those with undiagnosed IFNAR1 deficiency.

People with IFNAR1 deficiency can safely receive the rotavirus vaccine and non-live vaccinations, including the influenza and COVID-19 vaccines.

Healthcare providers need to be aware that children of Western Polynesian heritage who present for medical attention and are very unwell in the 1-2 weeks following MMR vaccine may need further investigation by an immunologist to assess for an immune deficiency.

Family members of individuals who have had a severe reaction to a live-attenuated virus vaccine, or are related to someone with known IFNAR1 deficiency, should be referred to an immunologist for consideration of the MMR vaccine prior to vaccination.

Dear Colleague,

I am writing to provide an important update regarding the MMR and MMRV vaccines, which are available under the National Immunisation Program for all children at 12 and 18 months of age respectively.

IFNAR1 deficiency is a newly described specific immune deficiency associated with severe adverse events and death following vaccination with some live attenuated virus vaccines, including the MMR vaccine, the yellow fever virus vaccine, and potentially the live varicella vaccine, though disseminated varicella post vaccination has not been seen.

A recent study from New Zealand and Australia (Attachment A) described 7 cases of children who were found to have this specific immune deficiency presenting for medical attention with a hyperinflammatory symptom complex of fever, rash, shock, and hepatosplenomegaly, or symptoms of encephalopathy within 1 week of MMR vaccination.

The research study can be accessed here:

<https://rupress.org/jem/article/219/6/e20220028/213170/A-loss-of-function-IFNAR1-allele-in-Polynesia> .

Although the role of MMR vaccine is not clear in all cases, 4 out of the 7 children in the study died and 3 had significant ongoing neurodevelopmental morbidity. An earlier report described complications following yellow fever virus vaccination in an adolescent with the condition. No deaths of children have been reported in Australia with IFNAR1 deficiency.

Although extremely rare, affecting less than one in one billion people, IFNAR1 deficiency appears more common in people who have 2 parents of Tongan, Samoan, or Niuean heritage. It is estimated to affect 1 in every 6,450 people with parents of Samoan heritage. This roughly equates to one child born every 2 years in Australia.

ATAGI has prepared a statement on IFNAR1 deficiency (Attachment B). The ATAGI statement does not recommend any changes to the National Immunisation Program.

ATAGI recommends that all people in Australia, including people of Tongan, Samoan, and Niuean heritage, continue to receive the MMR at 12 months of age and the MMR-V vaccine at 18 months of age, given that illness from wild-type measles and mumps infections is more severe in unvaccinated individuals, including those with undiagnosed IFNAR1 deficiency. People with IFNAR1 deficiency can safely receive the rotavirus vaccine and non-live vaccinations, including the influenza and COVID-19 vaccines.

Immunisation against measles is a key objective for the health and wellbeing of all Australians. Please ensure you remain aware of the advice contained in the ATAGI statement when considering administering the MMR and MMRV vaccine for your patient.

Yours sincerely



Dr Sonya Bennett
Acting Chief Medical Officer
29 April 2022

Attachments

Attachment A: ATAGI Statement: IFNAR1 Deficiency and Serious Adverse Events Following Immunisation