COVERAGE ALERT

New ICD-10-CM Diagnosis Code

Activated PI3K Delta Syndrome (D81.82)

EFFECTIVE OCTOBER 1, 2022

A new diagnosis code for **Activated PI3K Delta Syndrome (APDS)** has been established:

New! Diagnosis Code	Description
D81.82	**Activated PI3K Delta Syndrome (APDS)**

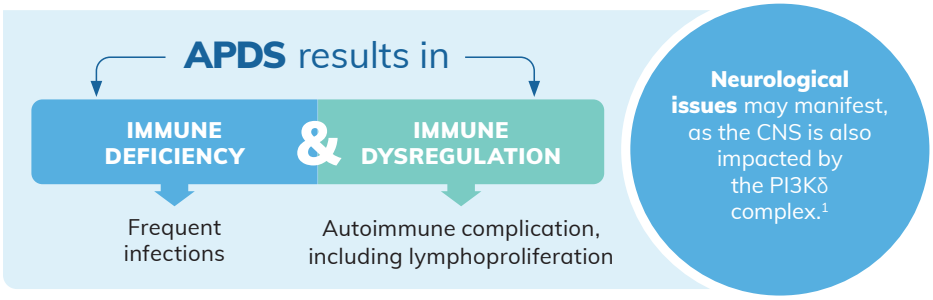
The information herein is provided for educational purposes only. It is the responsibility of the health care provider to select the proper codes and ensure the accuracy of all statements used in seeking coverage and reimbursement for an individual patient.

All about **APDS**

COMPLEX AND RARE

APDS is a disease of immune deficiency and immune dysregulation[1]

Characterized in 2013, APDS is a rare PI that affects nearly 1-2 people per million. This complex PI is caused by variants in the PI3Kδ enzyme complex (encoded by *PIK3CD* and *PIK3R1* genes), affecting both B cells and T cells.[1-4]



Pharming

All about **APDS**

COMMON SIGNS AND SYMPTOMS SEEN IN PATIENTS WITH APDS[5]

While symptoms can vary, APDS should be considered in patients with two or more of the following:

- Recurrent, severe sinopulmonary tract infections
- Persistent swollen or enlarged lymph nodes
- An enlarged spleen or liver
- Bronchiectasis (permanent lung damage)
- Chronic herpes virus infections
- Enteropathy (digestive issues, eg, diarrhea, abdominal pain or cramping)
- Autoimmune or autoinflammatory conditions (eg, cytopenias, arthritis, and eczema)
- Developmental delay (speech and growth)
- Lymphoma (cancer of the immune system)

For patients with two or more of the above signs and symptoms, a PI such as APDS can be screened for with a PI genetic testing panel.

DESIGNED TO ELIMINATE BARRIERS TO GENETIC TESTING

Genetic testing may improve both time to complete diagnosis and patient outcomes[6-8]

APDS has an autosomal dominant inheritance pattern that can be difficult to trace solely based on clinical history, as symptoms can vary even within the same family.[1,9,10]

If a parent is effected by the PI3Kδ variant, the risk of having a child with the **genetic variant is 50%**[10]

Disease management was changed in **40% of patients with a PI** after reaching a genetic diagnosis[7]

DESIGNED TO BE SIMPLE AND DEFINITIVE

navigateAPDS, a genetic testing program

navigateAPDS offers no-charge genetic testing and counseling for individuals who meet specific criteria. This program is sponsored by Pharming N.V., in partnership with Invitae.

NO CHARGE	No charge to qualified patients in the US and Canada
FAST	Receive results within 2-3 weeks on average (10-21 days)
SIMPLE	All forms for providers are available online. Blood draw kits, buccal swab kits, and saliva kits are provided
COMPREHENSIVE	A PI panel with over 400 genes is available
SUPPORTIVE	Free genetic counseling provided by GeneMatters

Academic immunologists and genetic counselors are working with navigateAPDS to help support you and your patients.

Visit www.navigateAPDS.com to learn more.

For more information on APDS, please visit **AllAboutAPDS-hcp.com**

References: 1. Coulter TI, et al. *J Allergy Clin Immunol.* 2017;139(2):597-606.e4. **2.** Jamee M, et al. *Clin Rev Allergy Immunol.* 2020;59(3):323-333. **3.** Lucas CL, et al. *Nat Immunol.* 2014;15(1):88-97. **4.** Orphanet. Accessed December 17, 2021. https://www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=EN&Expert=397596 **5.** Nunes-Santos CJ, Uzel G, Rosenzweig SD. *J Allergy Clin Immunol.* 2019;143(5):1676-1687. **6.** Sawyer SL, et al. *Clin Genet.* 2016;89(3):275-284. **7.** Quinn J, et al. *Immunol Res.* 2020;68(3):126-134. **8.** Chinn IK, et al. *J Allergy Clin Immunol.* 2020;145(1):46-69. **9.** Immune Deficiency Foundation 2007 National Patient Survey. **10.** Genetic Alliance; The New York-Mid-Atlantic Consortium for Genetic and Newborn Screening Services. Understanding Genetics: A New York, Mid-Atlantic Guide for Patients and Health Professionals. Washington, DC: Genetic Alliance; July 8, 2009.

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