Adagen is a (modified) enzyme replacement for adenosine deaminase deficiency. Adenosine deaminase is an enzyme that catalyzes the deamination of both adenosine and deoxyadenosine. Hereditary lack of adenosine deaminase activity results in severe combined immunodeficiency disease, a fatal disorder of infancy characterized by profound defects of both cellular and humoral immunity. It is estimated that 25% of patients with the autosomal recessive form of severe combined immunodeficiency lack adenosine deaminase.

**Pre-Authorization Criteria:**
as enzyme replacement therapy for adenosine deaminase (ADA) deficiency in patients with severe combined immunodeficiency disease (SCID) who are not candidates for or who have failed bone marrow transplant.

VCHCP requires Adagen to be prescribed by, or in consultation with, a physician who specializes in the condition being treated, due to the specialized skills required for evaluation and diagnosis of these patients.

**Dosing: Adult:**
Adult dosing is currently unavailable or not applicable for this drug.

**Dosing: Pediatric:**
- Infants and Children: Dose given every 7 days, 10 units/kg the first dose, 15 units/kg the second dose, and 20 units/kg the third dose; maintenance dose: 20 units/kg/week is recommended depending on patient’s ADA level; maximum single dose: 30 units/kg. All doses I.M.

Note: Treatment should be monitored by measuring plasma ADSA activity and red blood cell dATP levels (see product literature).
How Supplied:

Single-use vials, 375 units/1.5 mL; boxes of 4 vials

**Precautions:** headache; injection site pain; hemolytic anemia; thrombocytopenia.

**Drug Interactions:** pegloticase diminishes action of Adagen; reciprocal inhibitions with pentostatin

**References:**

2. [www.epocrates.com](http://www.epocrates.com): Adagen Drug information (Sigma-Tau Pharmaceuticals; June 2014.)

**Revision History:**

- Date Approved by P&T Committee: 1/28/14
- Date Reviewed/Updated: 1/8/15 by C. Sanders, MD
- Date Approved by P&T Committee: 1/27/15
- Date Reviewed/Updated: 1/13/15 by C. Sanders, MD; R. Sterling, MD
- Date Approved by P&T Committee: 1/26/16
- Date Reviewed/No Updates: 1/24/17 by C. Sanders, MD; R. Sterling, MD
- Date Approved by P&T Committee: 1/24/17
- Date Reviewed/No Updates: 1/23/18 by C. Sanders, MD; R. Sterling, MD
- Date Approved by P&T Committee: 1/23/18
- Date Reviewed/No Updates: 1/22/19 by C. Sanders, MD; R. Sterling, MD
- Date Approved by P&T Committee: 1/22/19
- Date Reviewed/No Updates: 2/18/20 by H. Taekman, MD; R. Sterling, MD
- Date Approved by P&T Committee: 2/18/20

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