

**A-T Clinical Center
Johns Hopkins Children's Center**

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Management of Acute Infection

A-T patients are predisposed to upper and lower respiratory tract infections because of immunodeficiency, aspiration, and impaired cough.

- Use of antibiotics should be considered for treatment of upper or lower respiratory tract infections that are severe, accompanied by fever, or persist for greater than 7 days.
- Precautions should be taken to reduce the risk of aspiration during respiratory illnesses.

Preoperative Evaluation and Surgery

A preoperative evaluation of lung function should be performed in all A-T patients regardless of age and whether or not they have chronic respiratory symptoms.

- Children and adults with A-T may have difficulty coming off the ventilator after surgery or other procedures requiring general anesthesia.
- Possible alternatives to general anesthesia and strategies that maximize airway clearance following anesthesia should be considered.
- Following placement of a gastrostomy tube, enteral feedings should be advanced with caution.



For more information, physicians can log in at [UpToDate.com](https://www.upToDate.com)

[atcp.org](https://www.atcp.org) | Tel: 800.5.HELPA-T or 954.481.6611 | info@atcp.org

QUICK INFORMATION FOR THE CARE OF PATIENTS WITH ATAXIA-TELANGIECTASIA

NAME

DATE OF BIRTH

Ataxia-telangiectasia (A-T) is an autosomal recessive disease characterized by:

- progressive neurologic degeneration with ataxia, eye movement abnormalities, dysarthria and impaired chewing and swallowing,
- immunodeficiency with lymphopenia and hypogammaglobulinemia of varying severity,
 - I have antibody deficiency and receive gammaglobulin therapy
- cancer predisposition (non-Hodgkin's lymphoma and leukemia most common),
- telangiectasia (often present), especially over the sclerae. In rare instances, telangiectasia may cause internal complications of bladder, brain, liver and lung.

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Diagnostic X-Rays and CT Scans in A-T

People with A-T have an increased sensitivity to ionizing radiation (x-rays and gamma rays).

- X-rays should be performed only when the result will affect medical management.
- If a patient with A-T has fever, cough, and breath sounds characteristic of pneumonia, a diagnosis of pneumonia can be made clinically and antibiotics can be prescribed without x-ray confirmation. If symptoms persist despite antibiotics, a follow-up chest x-ray may be useful.
- Routine screening dental x-rays should be avoided, but an x-ray to evaluate tooth pain is reasonable.
- In order to keep radiation to a minimum, patients should receive frontal view chest radiographs or radiation-sparing techniques for CT (e.g. fewer CT images per scan).
- There is no contraindication to MRI or ultrasound investigation.