

CASE TEACHING NOTES *for* "A Case of X-linked Agammaglobulinemia"

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INTRODUCTION

X-linked agammaglobulinemia is a rare disorder in which the affected individual fails to produce normal numbers of B-lymphocytes and therefore suffers from a severe deficiency of circulating immunoglobulin. Nonspecific body defenses and cellular immunity mediated by T-lymphocytes are unaffected. Children born with this condition typically experience recurrent bouts of infectious disease beginning at a few months of age. Once the condition is diagnosed, the individual must receive intravenous injections of gammaglobulin at regular intervals (usually monthly) throughout life.

Objectives

Upon completion of the case, students will have learned the following:

- The fundamental differences between nonspecific and specific body defense mechanisms.
- The differences between primary and secondary lymphoid organs, and the steps involved in lymphocyte "maturation."
- The distinction between humoral and cellular immunity.
- The nature of antigens, and why proteins are typically highly antigenic.
- The five classes of antibody and the means by which antibody molecules exert a protective effect.
- The differences between active and passive immunity.
- The role of maternal antibodies in the protection of an infant from infectious disease.
- The nature of X-linked inheritance.

This case study has been used in a sophomore-level course in human anatomy and physiology taught to premed and nursing students. In addition, it has been used in a senior-level elective course in general physiology taught primarily to pre-med students.

CLASSROOM MANAGEMENT

Students receive a printed copy of the case study after completion of the lectures covering the unit on the lymphatic system and immunity and at least one week prior to the class in which it will be discussed. Students are asked to review the concepts covered in the unit that are pertinent to the case, and then answer the questions to the best of their ability. Classes range in size from 20 to 30 students. Although I don't arrange the students in groups, I encourage them to work together in preparing the case. Reference materials are available on reserve in the college library, but most students use the internet as their primary source for answers to the questions.

Students are not required to turn in written answers to the questions, but rather, are called upon at random to answer the questions during the class discussion. Often, the discussion of the answer to each question goes beyond the initial scope of the question. Interestingly, students who tend to not ask or answer questions

during the lecture portion of the course, are often the most participatory when discussing case studies. Also, I have never failed to learn something new from the students regarding a case, no matter how many times I have used a particular case study.

Questions concerning the material covered in the case study are included in the next regularly scheduled examination. The questions are in the form of short-answer essays, requiring a short paragraph to answer. Typically, three case studies will be covered on each 100 point exam, with one question per case study valued at 5 points each. Though they are not verbatim, the questions follow very closely those that the students were required to answer prior to the class discussion.

Answer Key

Answers to the questions posed in the case study are provided in a separate answer key to the case. Those answers are password-protected. To access the answers for this case, go to [the key](#). You will be prompted for a username and password. If you have not yet registered with us, you can see whether you are eligible for an account by reviewing our [password policy](#) and then apply online or write to answerkey@sciencecases.org.

REFERENCES

Print

Harrison, T.R., et al. 1998. *Harrison's Principles of Internal Medicine* (14th ed). New York: McGraw-Hill.

Internet

X-linked Inheritance; Jennifer Butt, JMU.

<http://www.ikm.jmu.edu/Buttsjl/ISAT493/X-linked/xlinkedinheritance.html>

Sex-linked and X-linked Neuromuscular Diseases; MDA Ask the Experts

<http://www.mda.org/disease/40list.html>

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