

ARTICLE

The Woman Born Without a Cerebellum: A Real-Life Case Adapted for Use in an Undergraduate Developmental and Systems Neuroscience Course

Jennifer Brielmaier

Psychology Department, George Mason University, Fairfax, VA 22033.

In 2014, the case of a 24-year-old woman who had just discovered she was born without a cerebellum made headlines around the world. The details of this case were combined with other published cases of cerebellar agenesis to create an active learning exercise for an undergraduate developmental and systems neuroscience course. By reading an intriguing narrative and answering questions in stages, students work together to apply and

extend their knowledge of brain development and cerebellar function. The case can be used to introduce new information in a “flipped classroom” setting or as an interactive exam review.

Key words: active learning; case-based learning; collaborative learning; cerebellum; motor systems; brain development

CONTEXT

Current evidence suggests that problem-based approaches to learning have the potential to increase student engagement and motivation (Hmelo-Silver, 2004). Case studies are a prominent example of problem-based learning, and give students an opportunity to apply information learned in or outside the classroom to collaboratively solve a “real-world” problem. The Woman Born Without a Cerebellum was adapted from published cases of complete cerebellar agenesis (Glickstein, 1994; Timmann et al., 2003; Titomanlio et al., 2005; Yu et al., 2015) for use in a junior-level undergraduate course in Developmental and Systems Neuroscience. Not only does the case make for a compelling story, but it also allows students to apply their knowledge of multiple course topics such as genetic and molecular factors controlling early brain development, motor systems, and neuropsychological testing. This case could be implemented in any classes that teach these topics at an introductory level, and could be especially useful in courses geared toward pre-med students with an interest in neuroscience or neurology.

The case narrative presents the patient’s symptoms and results of neuropsychological tests without revealing the anatomical origin of her problems. Rather than being asked to solve the case right away, students work in small groups to answer questions that are presented in stages. Groups are not given the Stage 2 questions until they have correctly answered Stage 1 questions, and so on. Stage 1 questions focus on defining key terms and symptoms or tests that may be unfamiliar to the students. In Stage 2 students are asked to determine which CNS structure failed to develop in the patient, and must explain how they ruled out alternative possibilities. Stage 3 questions ask students to provide potential molecular and/or genetic mechanisms for cerebellar agenesis and to provide examples of brain areas/pathways that may have compensated for the lack of a cerebellum. This format creates a bit of friendly competition among groups as they work to answer the questions the most quickly and

completely (as judged by the instructor). Groups that finished more quickly were urged to spread out among the other groups and act as “coaches” to help their fellow classmates answer the questions without actually providing the answers.

The course in which this case has been used meets twice per week for 1 hour and 15 minutes. Because motor systems are the third and final unit of the course, the case has been used as a fun activity during the final class period. The course generally has 35 students, and they work in groups of 4-5 to answer the questions provided. At the end of the period, we come back together as a class and go over key aspects of the case. Students are shown MRI images of complete cerebellar agenesis and are given the opportunity to answer follow-up questions. The Classroom Implementation notes and full case narrative are available upon request from the corresponding author or from cases.at.june@gmail.com.

Learning Objectives

Content Objectives

At the end of this case, students will be able to:

- Define the terms dysmetria, dysarthria, nystagmus and Romberg's sign.
- Describe specific neurological tests that can be carried out to characterize deficits in motor and cognitive functions.
- Use knowledge of motor systems to identify which CNS structure failed to develop in a patient displaying specific motor symptoms.
- Describe the role of the cerebellum in cognitive functions.
- Compare and contrast the roles of different brain areas and motor pathways in producing reflex and voluntary movements.
- Apply knowledge of molecular and genetic processes involved in early brain development to identify possible mechanisms underlying the patient's condition.
- Apply knowledge of brain development and plasticity to

understand why the patient's condition might have gone undetected until the age of 24.

Process Objectives

- Locate and evaluate scientific information on the web
- Provide support for conclusions
- Hone collaborative problem-solving skills

CASE EVALUATION

Assessment

Because the case activity has been used during the last class period of the semester, the primary form of assessment has consisted of a few questions on the final exam. A total of three questions on the Spring 2016 final were deemed most relevant to the learning objectives of the case. The questions were a mix of multiple choice (1 point), fill in the blank (4 points), and short answer (4 points) for a total of 9 points. Sample exam questions can be found in the Classroom Implementation notes.

Scores for these questions ($n = 35$ for each) were compared to three "control" questions not relevant to the case but of similar Bloom's taxonomy level (Remembering). The score comparisons (case-relevant vs. control) were as follows: 31/35 (88%) correct vs. 23/35 (66%) correct for multiple choice; average score 2.8 vs. 3.4 for fill in the blank (statistically significant difference at the $p < 0.05$ level); average score 2.8 vs. 2.9 for short answer (no statistically significant difference).

Taken together, these scores suggest that students learned and retained information about the normal cerebellar function and the consequences of cerebellar agenesis as a result of completing the case study activity. However, it cannot be concluded that performance on case-related exam questions is significantly better than that for those not relevant to the case. In fact, students performed significantly better on the control fill in the blank question than they did the case-relevant question of the same format. It is possible that other differences would be seen with other methods of assessment.

Student Feedback

At the end of the class period, students in the Spring 2016 course ($n = 33$) rated their enjoyment of the case study

activity, the extent to which it enhanced their learning of course material, and their interest in working on case studies as part of a flipped classroom format. A 5-point Likert scale was used where 1 = Strongly Disagree and 5 = Strongly Agree. As can be seen in Table 1, students found the case study to be enjoyable and useful for enhancing learning. They were less enthusiastic about the possibility of replacing traditional lecturing with case study activities, perhaps due to lack of experience with this format.

SUMMARY AND FUTURE DIRECTIONS

This case is still very much in the pilot stage, having been implemented in the classroom only twice (and only once with assessment in mind). So far, however, it appears to be a useful activity for reinforcing previously learned information while giving students an opportunity to apply and extend their knowledge of brain development and motor systems. Students seem to enjoy the activity as a change from the usual lecturing, and it gives them a chance to develop skills in collaborative "real-life" problem solving. Preliminary data indicate that students performed well on exam questions related to the case, suggesting that case studies could be useful as interactive review sessions prior to exams.

In the future, the case may be modified so that student-generated rather than instructor-generated questions are used. Rather than asking students to define certain key terms, for example, they could be asked to generate their own lists. This change would incorporate a key feature of the Investigative Case-Based Learning format (Bioquest, 2016) as used by Roesch and Frenzel (2016) for Nora's Medulla. Such a model puts the inquiry into the hands of the students and allows them to practice the important skill of identifying what they do not yet know.

Another possible modification includes making the case narrative more interesting through the addition of "quotes" from the patient, her family members, or her examining physicians, and/or role-playing where students are assigned to play one of the "characters" mentioned above and act out scenes. It could also be useful to include additional assessments, such as classroom assessment techniques (e.g., "minute paper" or the clearest point/muddiest point exercise), or short presentations or

Survey Question	Average Rating (Mean \pm SD)	Example Comments
"I enjoyed the case study activity."	4.19 \pm 0.74	"I thought it did a really great job of enhancing my understanding of this topic and applying information to real life."
"The case study complemented my learning of the course material."	4.50 \pm 0.67	"This activity helped me to grasp the 'big picture.' It was especially good before the final."
"I would prefer to view lectures online before class and spend class time doing activities such as these."	3.78 \pm 1.26	"Learning with my group made me think about other possibilities that I would not have thought of. It was an interesting case study, and I enjoyed it."

Table 1. Students rated the case study activity as enjoyable and useful for enhancing their learning. A 5-point Likert scale was used to gauge their impressions, where 1 = Strongly Disagree and 5 = Strongly Agree.

written papers. All of the aforementioned changes would require expanding the case study activity to at least two class periods. If more time is spent on the case, exam items could go beyond simple recall of the facts of the case and target the higher-order cognitive skills laid out in the learning objectives. No matter how it is utilized, it is hoped that this case adapted “from the headlines” serves as a useful resource for instructors wishing to delve into problem-based learning in their own neuroscience courses.

REFERENCES

- Bioquest, “About ICBL” <http://bioquest.org/icbl> (accessed 04 August 2016)
- Glickstein M (1994) Cerebellar agenesis. *Brain* 117:1209–1212.
- Hmelo-Silver CE (2004) Problem-based learning: what and how do students learn? *Educ Psychol Rev* 16:235-266.
- Roesch LA, Frenzel K (2016) Nora’s Medulla: a problem based learning case for neuroscience fundamentals. *J Undergrad Neurosci Educ* 14:C1-C3.

- Timmann D, Dimitrova A, Hein-Kropp C, Wilhelm H, Dorfler A (2003) Cerebellar agenesis: clinical, neuropsychological and MR findings. *Neurocase* 5:402–413.
- Titomanlio L, Romano A, DelGiudice E. (2005) Cerebellar agenesis. *Neurology* 64:E21.
- Yu F, Jiang QJ, Sun XY, Zhang RW (2015) A new case of complete primary cerebellar agenesis: clinical and imaging findings in a living patient. *Brain* 138:e353.

Received August 15, 2016; revised September 26, 2016; accepted September 30, 2016.

The author thanks Dr. Kristen Frenzel, Dr. Leah Roesch, and members of the Neuroscience Case Study Development Group for helpful comments on the design of this case study exercise.

Address correspondence to: Dr. Jennifer Brielmaier, Psychology Department, 4400 University Drive, George Mason University, Fairfax, VA 22033. Email: jbrielma@gmu.edu

Copyright © 2016 Faculty for Undergraduate Neuroscience
www.funjournal.org