

UNDERSTANDING HUNGER & BARDET-BIEDL SYNDROME (BBS)

GABE'S STORY



A book for children and families living with BBS

Rhythm[®]
PHARMACEUTICALS

About Bardet-Biedl Syndrome

Bardet-Biedl syndrome (BBS) is a rare genetic disease that is estimated to affect about 1,500 to 2,500 people in the United States.

BBS is characterized by a wide range of manifestations, and everyone's experience can be different. Symptoms and severity can vary from person to person and may include extra fingers or toes (polydactyly), visual impairment, learning disabilities, kidney problems, intense, hard-to-control hunger called hyperphagia (hai-pr-fay-jee-uh), obesity, and other symptoms.

BBS is caused by variants, or changes, in a BBS gene that is passed down from parent to child. One copy of the changed BBS gene is passed down from the mother and the other copy is passed down from the father. In addition to the other symptoms, changes in BBS genes can stop a key part of the brain that controls hunger from working properly.

In someone living with BBS, the signals that turn hunger "off" don't work correctly in the brain. This can lead to intense, hard-to-control hunger and early onset obesity. Symptoms of intense, hard-to-control hunger may include taking a longer time to feel full while eating, feeling hungry again right after a meal, thinking about food constantly, and food-seeking behavior (sneaking or stealing food).

Gabe is our main character. He is 10 years old and has BBS. Gabe lives with his mom, dad, and younger sister, Melanie.

This is Hunger. Because of BBS, Gabe feels like Hunger is always with him. This hunger is intense and hard to control, which is different from how his family and friends feel hunger.



Gabe's story is intended to help children living with BBS make sense of their feelings, provide education on why they are experiencing intense, hard-to-control hunger, and recognize they are not alone.

Developed in collaboration with



Bardet Biedl Syndrome Foundation
& FAMILY ASSOCIATION

www.bardetbiedl.org

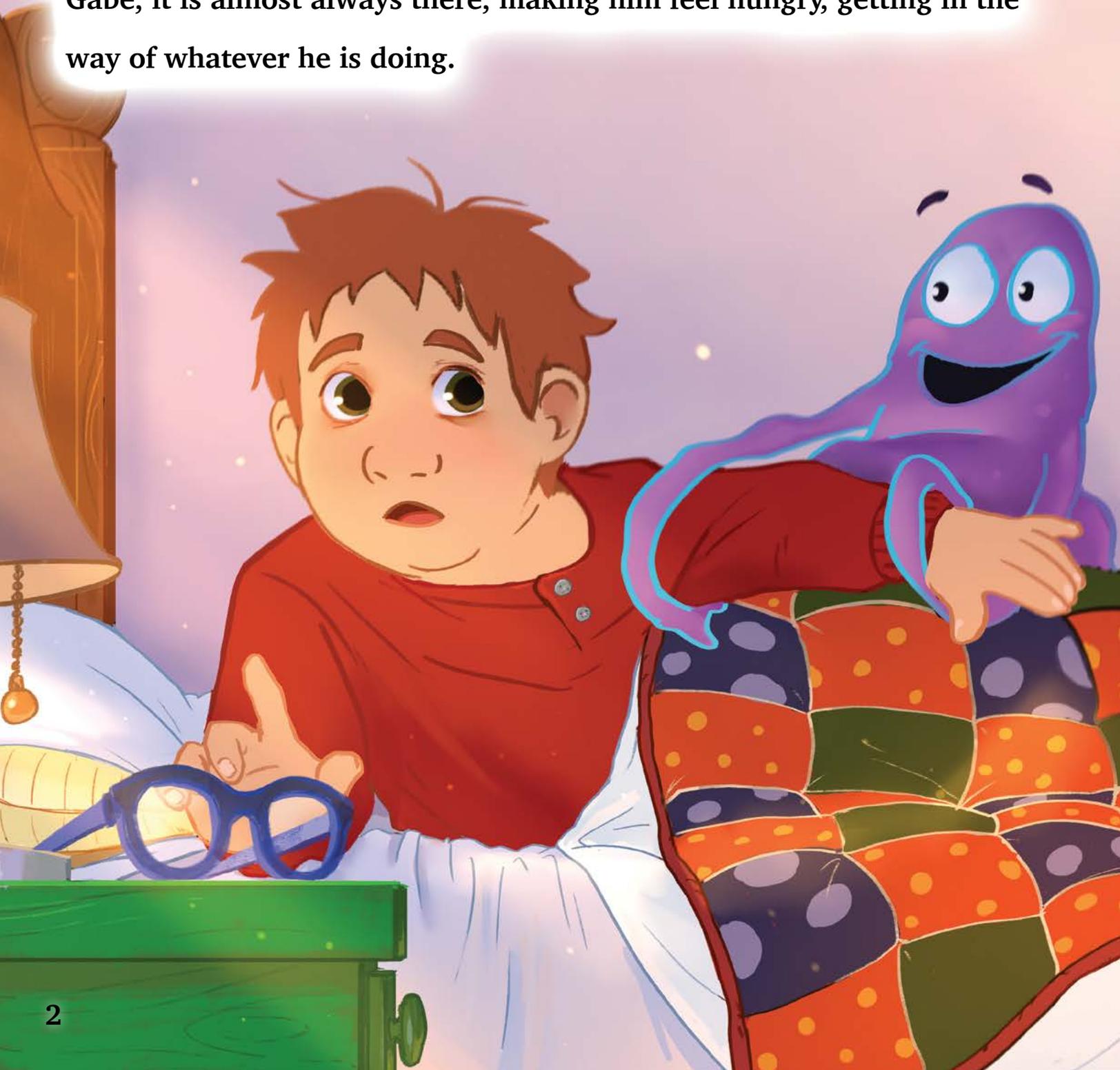


“Sun’s up, breakfast time! Get up, Gabe, get up!” yelled Hunger excitedly, bouncing on Gabe’s tummy.

Gabe was used to this morning wake-up routine. Hunger has been a part of his life for as long as he can remember. From the moment he opened his eyes, Hunger was there!

What Hunger feels like to Gabe is different from what other people feel. He has Bardet-Biedl syndrome, or BBS for short. It's a rare genetic disease that he was born with.

Intense, hard-to-control Hunger is just one part of living with BBS. For Gabe, it is almost always there, making him feel hungry, getting in the way of whatever he is doing.



“Good morning, Gabe! Rise and shine!” said Mom, coming into his room. **“Warm and sunny today, perfect for a trip to the park after school!”**

Gabe wasn’t thinking about the school or the park. He was feeling hungry.

“What’s for breakfast, Mom?” asked Gabe.



“Oatmeal, pancakes, and fruit,” Mom said cheerily as she helped Gabe pick out his clothes. **“It’s all ready for you. But first, let me help you get dressed and brush your teeth.”**

Melanie, Gabe's sister, was already at the table. "Mom made melon balls!" she said excitedly. Gabe loved fruit cut into fun shapes.

He ate his whole breakfast before Melanie finished her first pancake. Hunger poked him in the stomach, "Ask Melanie for hers! She never eats it all anyway! Ask, Gabe, ask!"



Gabe asked Melanie for her second pancake. Then Mom stepped in.

“Gabe, I know it can be hard sometimes,” Mom explained. “Your body has all it needs right now. You will have a snack and lunch at school.”



“Snack time is a long way away!” Hunger whispered in Gabe’s ear.

“What are you packing for snack and lunch? What about dinner tonight, Mom? What are you cooking?” asked Gabe.

“I’ve packed you hummus, carrots, and pretzels for snack. For lunch, a ham and cheese wrap, an apple, and veggie sticks with ranch dressing. After school, we’ll have celery and peanut butter. Then for dinner we’re having chicken on the grill, broccoli, and rice,” she said. “Now, time to get in the car!”

Gabe loved watching cartoons on his tablet on his way to school. But Hunger wouldn't stop bothering him. "So hungry.... belly hurts.... Ask for food. Now, Gabe, NOW!"



Gabe tried to ignore it. He turned up the volume. He squeezed his eyes shut. Then he couldn't ignore it any longer.

“MOOOOOOM, I’M SO HUNGRY! Can we stop at Fast Burger?!” Gabe cried. He knew he wasn’t supposed to yell like that, but he couldn’t help it.

“Gabe,” Mom said calmly. “Snack time is coming up at school. I packed you some carrots and hummus and…”



But it was too late. Gabe was too upset and frustrated. He started to cry. Melanie started to cry, too.

Mom pulled into the school parking lot and parked the car. She turned around to Gabe and Melanie. “Gabe, I hear you and I understand how you feel,” she said.

“Stop saying that!” Gabe cried. “You don’t feel hungry all the time! Melanie doesn’t feel hungry all the time.” No one understands, thought Gabe. It made him feel sad, frustrated, and lonely.

“What’s to understand?” Hunger nudged him.

“When you’re hungry, you eat.
Simple as that!”

As Gabe started to get out of the car, Mom said, “Wait, I have an idea. Melanie, can you give me your sketch pad and colored pencils?” asked Mom. Melanie wiped her tears and reached into her backpack.

“Remember what we learned from the doctors at the conference we went to last summer where we met other kids with BBS?” asked Mom. Gabe had been so excited to meet and hang out with other kids like him that he’d been distracted!

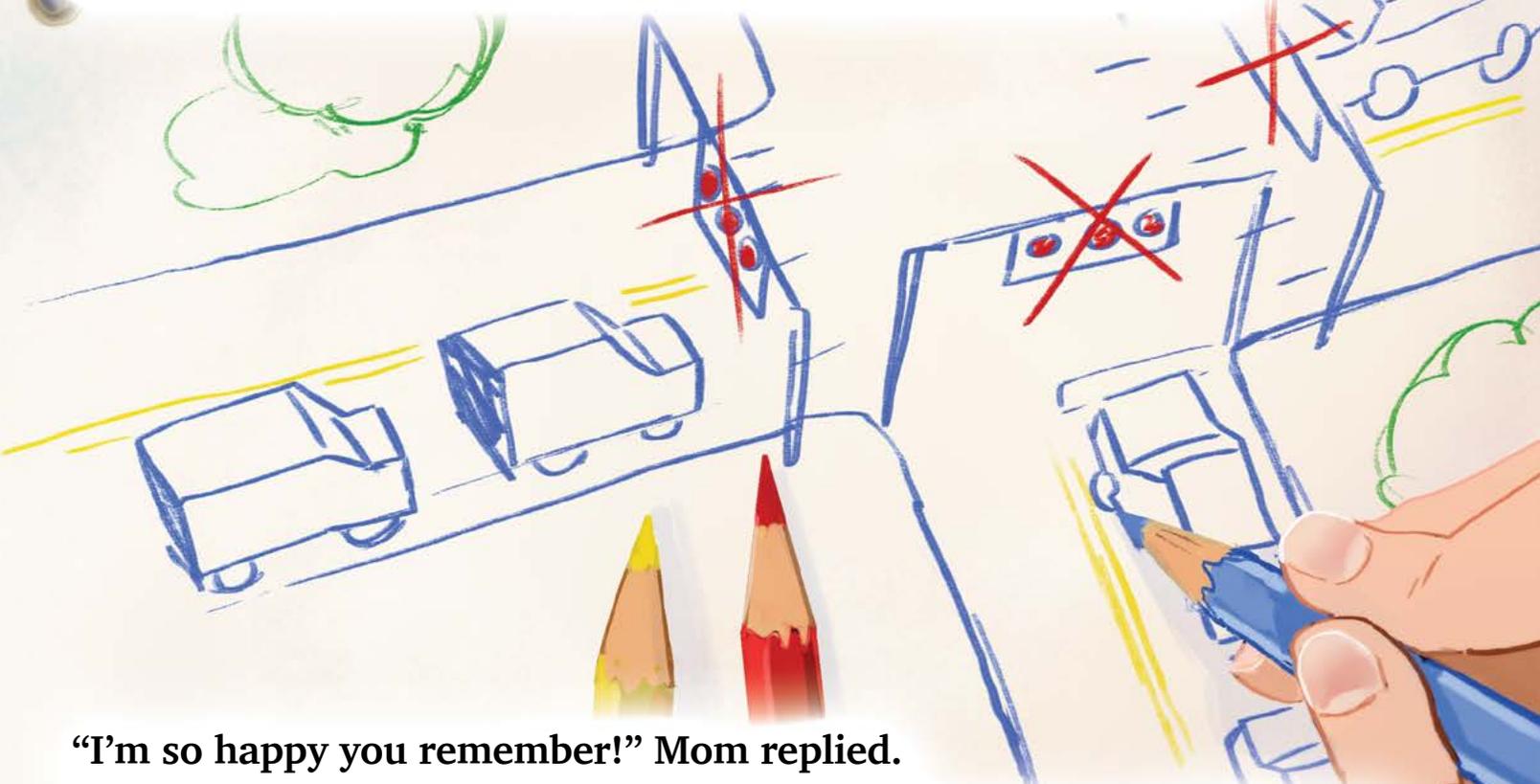
“Feeling hungry has to do with your brain,” said Mom. She drew a car.



“Think of your brain as a neighborhood full of roads. Cars carrying hunger messages travel between your body and your brain. A traffic light helps guide the cars to deliver their messages.”

As his mom drew cars driving past a traffic light, Gabe remembered.

“But because of BBS, not hungry messages in MY brain are stuck because a traffic light is broken,” said Gabe. “Even though my tummy is full, my brain doesn’t KNOW it because the messages can’t get through.”



“I’m so happy you remember!” Mom replied.

“When I get home, I’m going to hang this up next to our schedule as a reminder,” she added.

“But knowing WHY I am always hungry doesn’t make the feeling go away,” said Gabe. “And when I feel hungry, it makes me sad, frustrated, or even really mad.”

“That’s why I’m here to help you, sweetheart,” Mom replied. “And since your brain doesn’t know when it’s the right time to eat,” continued Mom, “it’s up to grownups who love you, like me and your dad, to help. We will always make sure your body gets enough food.”

Gabe smiled because he knew his parents would always take good care of him. “Thanks, Mom,” he said. He took a deep breath, opened the door, and headed into school.

In math class, everyone was working in groups sorting coins. “Come to our group, Gabe!” called Ava. “You can do the quarters!”



Gabe made perfect little stacks as he counted. While his group waited to hear what to do next, he felt that familiar hungry feeling, even though he had his morning snack. “The nurse’s office has crackers for sick kids!” Hunger demanded. “Ask to go to the nurse! Ask, Gabe, ask!”

Gabe was upset that Hunger kept distracting him. He was having fun working with his classmates!

Mrs. Cruz called Gabe to her desk and quietly asked him what was wrong. “I feel so hungry. It’s hard to pay attention. All I can think about is food,” whispered Gabe.

“It must be really hard to concentrate when you are feeling hungry,” she said. “Thank you for telling me that.”



Mrs. Cruz pointed to the daily schedule on the wall, which showed each class period in a different color. “We have 30 more minutes of math, then we’ll be lining up for lunch.”

She held up her pinky. “Can you try to focus on classwork until then?” Gabe really liked Mrs. Cruz, and pinky-promised to try his hardest.

At the end of the school day, Gabe was tired. He couldn't wait to get home. Hunger pestered him the whole bus ride. "Gabe, Gabe, after-school snack is next!"

Gabe was thinking about the peanut butter and celery that would be ready when he got home. He didn't notice they had reached the bus stop until Melanie called his name.



After his snack, Gabe asked about dinner time. “We’ll have dinner at 6 after Dad gets home, like always,” Mom replied.

Hunger didn’t like the sound of that! “We can’t wait that long!

Wait for Mom to go, then check the cabinets, OK? Check, Gabe, check!” When Mom turned around, Gabe thought about listening to Hunger and trying to find another snack, but then he looked at the whiteboard where Mom had put the picture of the cars. He took a deep breath, remembered what she had told him, and tried to stop thinking about how hungry he felt.



Then Mom reminded him it was time to meet his friend Owen at the park. Gabe loved playing with Owen! He made up the best adventures. Gabe was happy to have something else to think about to try to forget about his hunger.

Gabe and Owen set out on a mission collecting pine cones. They piled them under a big bush. “The aliens will never find them!” shouted Gabe, laughing.



Gabe found a huge pine cone and brought it to show Mom. “Is it time to go home for dinner yet?” he asked. He was having fun, but thinking about dinner kept distracting him from the game.

“Gabe, come back! The alien invasion is starting!” Owen called out.

On the walk home, Gabe told Mom about the game. She smiled “What an adventure! I’m so proud of you for playing so nicely at the park.”

Gabe was going to ask about dinner when Dad called from the front door. “How was everyone’s day? Come on, dinner is ready!” he said with a smile.



Everyone did their part to get the table ready. Mom put out the dishes and silverware. Gabe filled the water glasses. Melanie placed a napkin at each plate. Dad brought the food to the table.

Gabe ate everything on his plate, but it wasn't enough to keep Hunger from nagging him! "I'm still hungry. Mom and Dad, may I please have some more?"



Mom smiled, "Thank you for calmly telling us that you are still hungry and asking politely for more. You can have one more scoop of broccoli. That will be enough for your tummy until popcorn and TV time."

Gabe felt proud for being calm when he asked for more food. He looked at the picture hanging on the whiteboard and remembered what Mom had said about his hunger messages getting stuck because of the broken traffic light.

When Dad came in to say good night, Gabe was sitting on his bed. Hunger was stretched out, blocking him from lying down! “Can I have one last snack, Dad? Please? I promise to go right to sleep.” Dad reminded Gabe that his tummy was already full. “Remember the delicious dinner we had? And the popcorn afterwards? When you wake up, scrambled eggs and toast will be ready for you.”

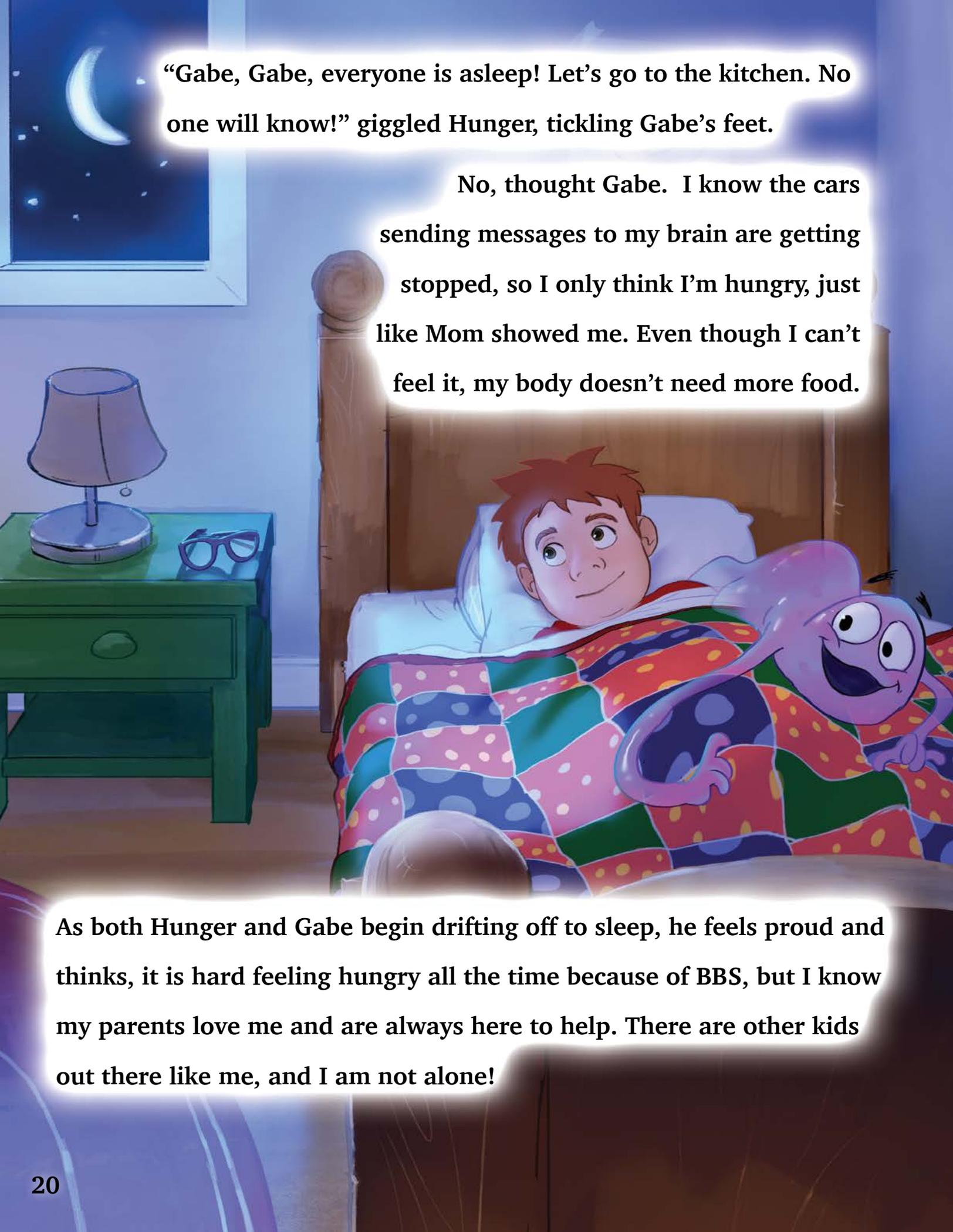


Dad put his hands up for their special high-five. I'll get to eat in the morning, thought Gabe, just like Dad said. Mom and Dad always keep their promises.

Mom came to say goodnight. “Your tummy is full, your teeth are brushed, just one thing left!” she said. Gabe reached up for a hug. “I love you, Mom. Good night!”



“I love you too, sweetheart. Good night!” said Mom, leaning down and planting a long kiss on his forehead.



“Gabe, Gabe, everyone is asleep! Let’s go to the kitchen. No one will know!” giggled Hunger, tickling Gabe’s feet.

No, thought Gabe. I know the cars sending messages to my brain are getting stopped, so I only think I’m hungry, just like Mom showed me. Even though I can’t feel it, my body doesn’t need more food.

As both Hunger and Gabe begin drifting off to sleep, he feels proud and thinks, it is hard feeling hungry all the time because of BBS, but I know my parents love me and are always here to help. There are other kids out there like me, and I am not alone!

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Rhythm InTune is a support program designed for caregivers and people living with rare genetic diseases, including BBS.

If you're interested in speaking with a Patient Education Manager about the education Rhythm InTune can offer you or the person you care for, you can email or call us at:



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Rhythm Pharmaceuticals is a commercial-stage biopharmaceutical company committed to transforming the care of patients living with rare genetic diseases, including Bardet-Biedl syndrome (BBS).



The Bardet-Biedl Syndrome Foundation and Family Association is dedicated to improving the lives of individuals and families affected by Bardet-Biedl syndrome (BBS). Our mission is to provide a community of support for individuals and families; information about BBS; and promote science and research to improve the lives of individuals with BBS.

www.bardetbiedl.org

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