

More information about the diseases covered in this book: www:guiametabolica.org

The Metabolical Gang Collection

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Mysterious Fortprints

Hello, I'm **Annie**.

I love running, swimming and playing! When I grow up I am going to be a pilot. I have citrullinemia and I love pineapple so much. Oh, I'm 10 years old.

Well, I'm **Laura** and I like cooking with my parents and drawing princesses. I'm eight and a half years old.

I am ten years old and I can explain a lot of things about PKU and other stuff about metabolism. They know me at the library.

I'm **Gabriel**.

My name is **Arnau**. I am ten years old and I love to read. As I have homocystinuria, I can't eat any animal protein. On the contrary, I adore all living creatures.



My name is **Thomas**. I really like ratatouille, doing puzzles and jumping. I am four years old. I hate going to bed before the others.





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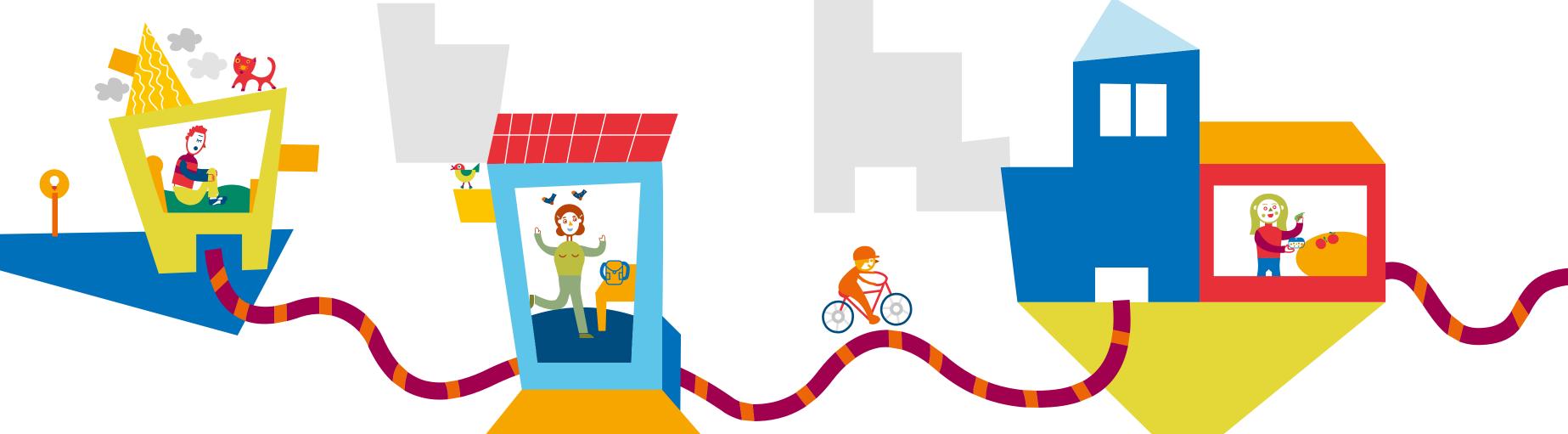


It is a grey day, like Arnau's mood.

The thick winter clouds seem to have plotted with his mood. His friends must be getting themselves ready to go on a camping trip, but he has decided not to go. They kept insisting that he go, but wrinkled his eyebrows and said no over and over again.

While Arnau idles in bed with thoughts as grey as the sky, Annie, a few blocks away, finishes filling her rucksack with her favourite socks, the ones with rockets and the ones with giraffes. She is more than happy: she is eager and jumping up and down all through the house, whistling and hoping that her father will finish preparing the bag with the food.

At the same time, nearby, Laura fills her lunchbox with magic meatballs and puts apples and pears in a bag. This will be her lunch for the first day, which each of the children has to bring. She is thoughtful and a little sad because her friend Arnau has decided not to go on the camp. Even after she had promised that she would bring a special dish to share with him. Laura and Gabriel were the ones who had most insisted that Arnau should come with them, but he told them that he didn't want to go camping with people who ate "weird stuff". He said it just like that, that he was tired of hearing about the camp and he preferred to stay at home playing with his computer, reading and painting animals from the book *Surprising Fauna* that he had got for Christmas.



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Gabriel is sorry that his friend is not going to camp. Many Saturdays they meet at the library to look for information about PKU or homocystinuria, which are the diseases they both have, and about other metabolic problems. Gabriel writes in his diary with language that is half technical, half infantile, describing things. Arnau does the illustrations. In the diary you can read things like this: "in the year 1983 Woo discovered the gene that manufactures phenylalanine hydroxylase" or like this other one: "in citrullinemia, ammonia is accumulated."

"Whaaat?"

Annie shows surprise on her face and her mouth opens in awe each time her friends leave the diary in her reach.

"What? What's this about ammunition?"

Arnau laughs and Gabriel explains that it is ammonia, not ammunition. It is the substance which his blood has in excess, above all, if he does not eat as he should.

"Leave me out of the bad words," Annie always says. She's not at all interested in her metabolism and would rather pretend to be an Amazon riding through the forest.





Yes, Annie is like a wild horse. She always finds some excuse to spend her time trotting, running, imagining that she is an astronaut or that she is diving in the South Seas or any other character that can't keep still for a second.

Laura is very willing, she signs up for everything. On the other hand, Gabriel quickly gets tired of running around and prefers to go back to his books and notebooks. When he turned nine, his aunt gave him a diary for him to write everything that came to his mind. And that was how he came to love scouring the library: he decided that he was going to know more than anyone about different congenital metabolic disorders of his friends. Later on, he convinced Arnau to do the drawings. Arnau's mind is more creative than investigative, but dragged along by Gabriel, he has become one of the library's most celebrated bookworms.

While his friends are preparing to go to camp or not to go, Thomas pulls at his socks and listens to his father's instructions once more:

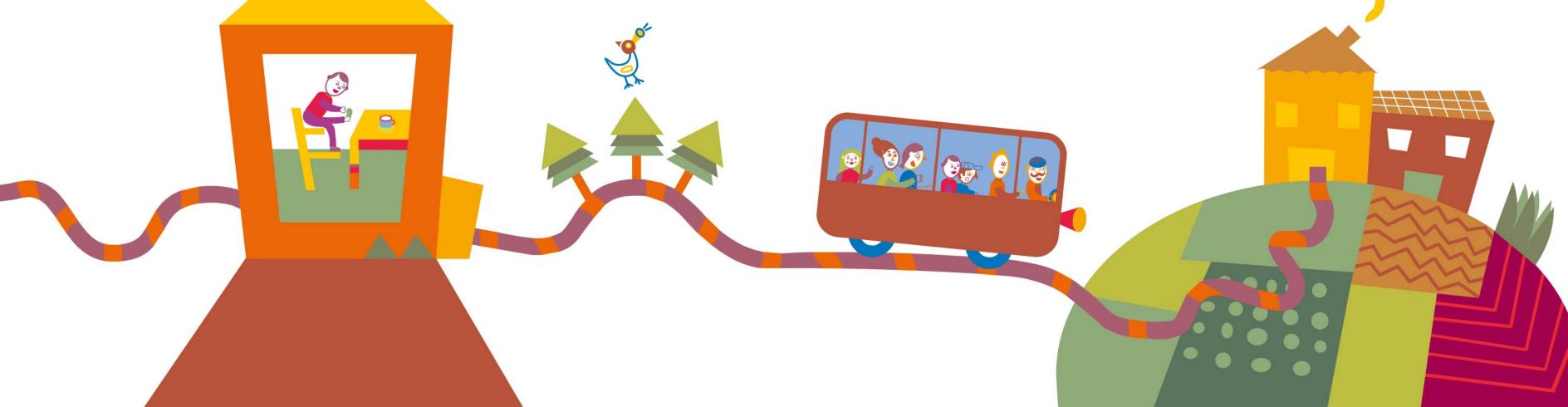
"You know that you have to eat up all of your food and listen to the monitors until mum gets there, right?"

He says yes, doing a little dance with his legs, eager at the prospect of going to camp. It is the first time that Thomas, who has leucinosis, will spend a night away from home. His mother will come at the weekend. She is a very good cook and knows a lot of recipes with little leucine. Tomas is lucky to have older and well-documented friends like Gabriel, Arnau and Laura. He doesn't have any problems with food: he likes whatever he is given and he is crazy about vegetables, above all, well sautéed with a little garlic!

Now everyone is ready to go. The journey to the hostel goes by quickly with songs and riddles. In total there are twelve children, five monitors and two family members: Annie's father and Gabriel's mother, loaded with boxes of food to spend these days together.

The house is at the top of a small hill surrounded by meadows, one hundred metres from a grove. When the cars arrive, one of the monitors has already taken charge of lighting the fire, and a smoke spiral coming from the chimney welcomes them. A dog from the nearest farm accompanies a flock a few metres away from the pastor.

"I am sure that we are going to have an adventure here," Annie says, before running up to choose her bed, beside the window. From there, she has a strategic view of the valley and the surrounding farms.



Gabriel, followed by other children, prefers to explore the kitchen, where the monitors are putting the food in the fridge and in hampers.

"My mother has taught me a new recipe," he says, showing his diary to the monitors. "She got it from a book in French."

The monitors take Gabriel's diary and read the title of the recipe: "Colonial Cake". They laugh.

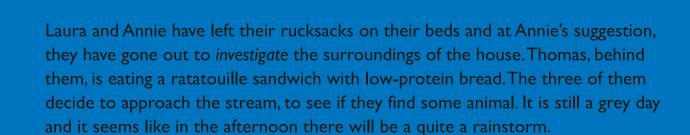
"So you want us to make a special cake for the end of camp?" asks the girl.

"Well, I'd prefer us to do it for the start," responds Gabriel with such an innocent face that he makes the monitors laugh even more.

"Don't worry, we'll make it sooner or later...if we have the ingredients."

But Gabriel, who had it all planned out says "We have brought them!"





Luckily, they have all brought books and games to while away the time inside the house. But while it's still dry, Annie wants to spend as much time as possible outdoors.





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"Hey look!" she points to some rocks above. "Something is moving. Do you see it?"

Laura looks towards the rocks and her heart beats heavily. Thomas smiles and jumps up and down, hoping that Annie will lead the expedition to the side of the river where it seems that something peculiar is happening.

Little by little, single file, the three approach the rocks, where they discover that the "something moving" is a bag hanging from a branch.

The first expedition has brought a result of a supermarket bag and little more: some small coloured stones in amusing shapes.

It is time to eat and everyone sits around a big wooden table in the kitchen with their delicacies. The one who takes the prize, without a doubt, is Laura, with her magic meatballs. They look great!

"Mmm! What are they made of?" asks Annie, without taking her eyes off them.

"They have mushrooms, potatoes, and sautéed onion, garlic and parsley."

"And YOU made them? You know all the ingredients!" Annie thinks that even washing a tomato is epic. In contrast, Laura is quite the chef. Gabriel's mother strokes her hair and says to Annie:

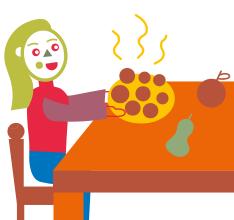
"Laura can show you how to cook the magic meatballs, right?"

Laura nods shyly. Nothing would make her happier than being the cookery teacher for her friend. For once, she would get to lead the game.

"Well if they make meatballs tomorrow, the rest of us can make the Colonial Cake, "Gabriel demands opportunistically. But his mother reminds him of what the monitors said, that the cake would be to celebrate the end of the camp. Thomas also puts in his request:

"And ratatouille sandwiches!"





After supper it is time for board games. Thomas's parents, who are graphic designers, have invented a game which a friend of theirs has constructed in wood. Each piece represents a food. In this way, Thomas

learns what he can and cannot eat. Today he makes a mistake with the shrimps and the almonds. Lucky that Gabriel and Laura are beside him to tell him where he goes wrong and when he is right.

"You got 18 right!" exclaims Laura the optimist.

"You got 2 wrong, "rules Gabriel, who usually sees the glass half empty.

While Thomas and another girl are occupied putting the pieces together again, Annie's father takes out some cards so that the older ones can play. He got them from a website. With these cards, apart from playing, they learn things about the correct nutrition of a girl or boy with a metabolic disorder. This is great for Annie, since she doesn't show much interest in the citrullinemia that she suffers from. In contrast, by playing the game, she remembers information that is important for her.

"Pineapple?...Mmm, yes, I can eat pineapple. Coconut?...Mmm, no, I can't eat coconut. Tangerines?...Mmm, of course I can. Papaya?...What is papaya?"

Gabriel takes advantage of the opportunity to show off his scientific knowledge.

"Thomas has Maple Syrup Urine Disease because his leucine accumulates."

"Whaaat?" If he takes the medicine syrup, he'll become a leek?" Annie jokes.

No need to say that when Annie and Gabriel put their minds to it, they make a great comedy duo.

The camp goes on with the rhythm of games, discoveries and shared moments. The wind and rain gave way to sunshine. The monitors have organised an outdoor game called *The conquest of the castle*. They create two teams and give each a colour. One monitor gives yellow and red handkerchiefs to the members of each team. They have to use them to cover their face and hair so that they cannot be recognised. The objective is for one member of one of the teams to conquer the castle, which consists of some rocks which have been made into a mound. The children and the monitors have to hide and find a way to reach the rocks without a member of the opposing team discovering them and saying their name out loud.

Everybody starts to run and search for a good hiding place. Gabriel is thinking of the best way to get to the castle without being seen. Laura is tying on her handkerchief when someone pulls her arm.

"Laura!" a familiar voice whispers in her ear, "Let's not waste more time on this absurd game. Come, I have found a very mysterious thing."

Laura doubts between following Annie into the forest and stays hidden, waiting for the moment when someone dares to venture out to conquer the castle.

"Come on, nobody will miss us for a while!"

In the end, she follows her because she likes to feel her own heart beat like a drum and her breathing speed up. They make their way, between elms and willows. Annie runs with gazelle leaps and Laura follows puffing, until they reach a small area of level ground from which they can hear the voices of the group in the distance.

"Look!"

"It's a bear!" she exclaims, putting her hands on her cheeks to exaggerate the sensation of surprise.

"No Laura, it's something worse..." Annie looks intently at her friend and puts on the voice of a private investigator, "this could be an extraterrestrial."

Annie's theory is that the footprints have a round form and the claws can't be distinguished, therefore it can't be an animal. It cannot be a person either since there are no marks of boots or shoes. Laura does not want to continue there, alone and far from the others, before such a strange discovery.

"We have to tell Gabriel!" She runs away, without waiting for Annie, who is trying to find another clue.

Laura runs and runs until she discovers that she is lost. A feeling of fear comes upon her in seconds. Then, a hand on her back forces her to utter a sharp scream of fright. "Laura, you are out!"

One of the children has found her and has taken off her red handkerchief, so she has lost the game. Happy to go back with the rest, Laura runs to look for Gabriel, who has been crowned winner. His powers of observation have brought him victory. Excited as he is, he pays no attention to Laura, until, at mealtime, someone asks for Annie,

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"Have you seen her?"

The monitors and, above all, Annie's father begin to look for her in the area around the house.



Laura approaches Gabriel's mother and explains, ashamed, about what happened during the game. Now she is sorry to have left her friend alone. Everyone gets into groups to look for her while Laura directs the group to the place where they found the footprints. But Annie is not there. The older ones organise themselves into three groups: one will go upriver, another will go downriver and the third will stay in the small area of level ground with the little ones, to see if Annie returns.

While the two expedition groups are out, Laura seizes the chance to explain about the footprints to Gabriel. Surreptitiously, she takes him to the patch of ground where they found the prints and whispering, she tells him about Annie's theory. Luckily, the disappearance does not last long. Annie was not too far away. She is as happy as a lark about her adventure. They see her appear with one of the groups of adults, without even a hint of fear on her face, even though her father has told her that later on they would have words, just him and her.

Night falls and everyone is playing or preparing activities for the next day. Annie has just had a conversation with her father, he told her about the dangers of breaking away from the group. He told her that he would not punish her, but he asked her to make a promise: if she is old enough to decide things on her own, she also has to be responsible for her food. She has promised him that she will be and Laura will teach her to cook magic meatballs. Her father has also made a promise...when the school year finishes, if Annie holds up on her side of the deal, they will go horseback riding.





Delighted, Annie enters the main room like a tornado. There, Thomas and some small girls are painting a poster. She goes to Laura and Gabriel. Her eyes light up when she tells them that she is going to go horseback riding. A dream going to come true!

But the other two are worried about something else.

"They were really strange, those footprints..."

Gabriel has decided to participate in the mystery and to guess who or what the footsteps come from. Because Annie's theory, the one of the extraterrestrial, doesn't convince him at all. Laura insists that it must be a bear and Gabriel tells her that for a bear, the prints are too small. But they are too big to belong to a squirrel.

Gabriel has a swig of Captain PKU juice while he thinks. Annie says to give it up, that it is a Martian who has surely gone home to his planet after all the commotion. So they won't be able to find him or to test her theory.

"We need a person who is an expert in animal prints."

"You think that your father..." Laura is talking to Annie but Gabriel interrupts her:

"Arnau! He knows heaps about animals! The other day at the library he spent the whole afternoon with a book about mammals."



"And what were you doing in the meantime?" Annie teased.

"Well, looking for information for a friend of mine who has galactosemia, Gabriel replies, purposely using one of the weird words that Annie always makes jokes about. 25

"You have a galactic friend!" She exclaims with her blue eyes as round as saucers.

"Yep, that's it!" Laura laughs. "But do you at least know what your disorder is called?"

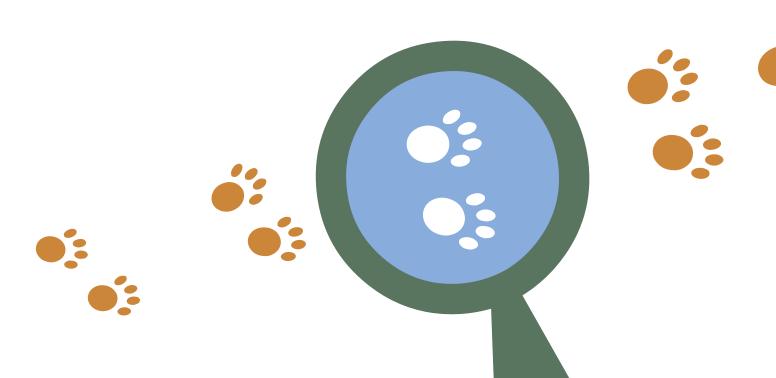
"Of course. I'm not stupid. It is called citrullinemia and it means that I have too much citrulline.

For once, Gabriel returns the joke:

"Must be because you eat too many citrus fruits, like tangerines!"

Everyone laughs, including Annie. But she reminds them of the matter to hand.

"Ok, ok, but you said that we need an expert in footprints and that Arnau could help us. But there is a small problem: Arnau hasn't come to camp."



The three of them agree and have asked Gabriel's mother to call Arnau. She is a friend of Arnau's mother, and she calls. She speaks to her friend for a moment and then she passes the telephone to her son.

"Arnau, we have found some footprints! We don't know where they came from. You have to come, we need you! Come and bring your book Surprising Fauna."

With this clear and convincing message, Gabriel has made Arnau want to join his friends.



The weekend has arrived. Thomas's mother has arrived at the hostel and she has brought with her a person that everyone was anxiously waiting for: Arnau. With his book under his arm and a big smile, he will spend the last days of camp with his friends.

They have given him just enough time to settle in and everyone has flown out to the small level area, including Thomas, who doesn't want to miss out. They want to investigate the footprints.



The footprints are still visible, even though the ground is already dry. Arnau looks at them, traces their shape with his fingertips and leafs through his book to the drawings of animal footprints. Everyone crowds around him, holding their breath.

"It's a dog. Without a doubt." He shows them photos and drawings of dog footprints that look very similar.

Gabriel starts to laugh. Laura speculates about whether it would be the dog from the neighbouring farm or an abandoned dog. Annie makes a funny face and goes back to the house after saying that surely it is an extraterrestrial dog.



Everyone is very happy, not just to have discovered the secret of the footprints, but because finally Arnau has come. Now he is the centre of attention and everyone wants to flip through his book of fauna. Annie is the only one in a bad mood.

"Did your father scold you much?" Laura asks, trying to console her.

"He didn't punish me. He wants me to learn some recipes and help him with the cooking."

"And that's why you're sad? Cooking is fun. You will see when we make the magic meatballs!"

"It's not that. It's just... well I am the one who found the prints!"

"That's true. We all know it. So also thanks to you that Arnau has come."

Annie realizes that her discovery, extraterrestrial or not, has been worthwhile.

She hugs her friend and proposes that they go and look for coral under the bunk beds.





Camp comes to an end. In the afternoon, everyone will pack away their things and they will go back to the city. But before, the big surprise: the Colonial Cake that they all baked together. Following the instructions from Gabriel's mother's recipe, some made the dough with low-protein flour, others measured out the ingredients: the cinnamon, the egg substitute, cane sugar...some cut the fruit to decorate the cake and the youngest set the table. Even Annie helped. She asked for them to let her stir until the dough was just the right consistency.

What a cake! Everyone is licking their fingers. The plates are clean and the children still have a little fruit syrup making them moustaches. Thomas has had seconds. Gabriel and Arnau are having a discussion about the quantity of phenylalanine and methionine the cake has.





Annie and Laura are singing and dancing a song they made up about Martian dogs and flying horses.

One thing that they are all sure about is that they will come back to camp. Arnau says that nothing in the world could ever make him miss it. Now he knows that he can be just fine when he is far from home. With or without homocystinuria, he can have all kinds of adventures. Like the one of discovering the mysterious footprints of the shepherd dog.



Inborn errors of metabolism: This is a very numerous group of "rare" diseases caused by hereditary DNA changes. These mutations cause proteins to function incorrectly, and cause an alteration in the metabolism because they interrupt one of its functioning pathways.

Some of these diseases, the ones that affect the metabolism of amino acids and organic acids, are treated with special diets which prevents the accumulation of toxic products which can affect the brain and other organs of the affected child.

This means a treatment based on diets low in foods with a high protein content (meat, fish, eggs, milk and derivatives) which are substituted by a special formula (Captain PKU juice in the story) without harmful substances for these children, but with everything else necessary for health.

PKU or Phenylketonuria: A congenital disorder in the metabolism of phenylalanine, which causes the accumulation of this substance in the blood, urine and tissues, causing harm to the brain in particular.

Homocystinuria: A congenital disorder in the metabolism of homocysteine, which causes its accumulation in plasma, urine and tissues, harming the brain, bones, eyes and the circulation of blood.

Ammonium: Proteins are formed by very long chains of amino acids which liberate ammonium when they are "broken down". Ammonium is a substance which is very toxic for the brain. Our organism eliminates it by converting it into urea, by means of a series of cyclic reactions. The urea cycle converts the toxic ammonium into urea, which is not toxic and can be easily eliminated in the urine.

Citrullinemia: Defect of the urea cycle which causes the accumulation of ammonia and citrulline in the blood, urine and the brain, which is particularly harmed by ammonia.

Leucinosis or Maple Syrup Urine Disease: A congenital metabolic disorder of some amino acids: leucine, isoleucine and valine, which causes the accumulation in plasma, urine and tissues of neurotoxic products, which smell like maple syrup (hence the name of the disease). Leucine and a derivative of it are the compounds which most accumulate and are also the most toxic. That is why this disease is

Low-protein bread: In general, it is bread without gluten. It is used in the treatment of congenital metabolic disorders of amino acids and organic acids, as is gluten-free pasta.

called leucinosis.

Galactosemia: Congenital metabolic disorder of galactose, a sugar contained in milk. It is treated by eliminating milk and its derivatives from the diet.





Other titles from the collection: The Secrets of the Mulberry Bush * A Prize Recipe

Everybody is different, inside and outside.

Some are dark and have long legs; others are blonde and chubby. Some have small ears and others, eyes like saucers. That is what you can see on the outside. But inside our bodies there are also differences. Some people cannot eat sugar, others cannot tolerate milk or lactic products; some have allergies or can't eat animal proteins. Luckily, just like there are clothes for every body type, there are foods for every type of metabolism.

This collection will introduce you to Annie, Arnau, Gabriel, Laura and Thomas, boys and girls with their individual characteristics, but who have in common a special metabolism. With them, their friends and their families, you can discover about phenylketonuria, galactosemia, maple syrup urine disease or citrullinemia, and with them you can have mysterious, funny and exciting moments.

With the Metabolical Gang you can lean a lot about disorders and nutrition and have fun with their adventures.





