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More research needed on hypothalamic syndrome

11-05-2022

A benign tumor in the hormone center in the brain - craniopharyngeal - can lead to what is known as hypothalamic syndrome. This condition disrupts the hormonal balance and has major consequences for quality of life. Dr. Hanneke van Santen has set up a multidisciplinary team with colleagues from the Prinses Máxima Center for Pediatric Oncology and the Wilhelmina Children's Hospital to guide children with hypothalamic syndrome. Van Santen: 'As a team, we are building expertise in relation to this rare, as yet uncurable condition.'

Hypothalamic syndrome is a rare disorder of the hormone center in the brain. In some children, the syndrome arises from a genetic predisposition, but in some it is caused by a tumor attached to the hypothalamus, an endocrine gland in the brain. Hypothalamic syndrome has major implications for growing children and their families because of the role the hypothalamus plays in controlling hormones and appetite, among other things. Tumors that can lead to hypothalamic syndrome are notably the craniopharyngeoma , but a so-called low-grade glioma or a germinoma can also grow in this location. The prognosis of these types of tumors is good. However, the quality of life can deteriorate significantly.

About the hypothalamic syndrome, Hanneke van Santen, pediatric endocrinologist - a physician specializing in hormones - at the Máxima and UMC Utrecht, wrote a review article with other international experts that recently appeared in the renowned scientific journal Nature reviews | Disease Primers.

Limiting neurosurgical damage

'The craniopharyngeoma is located in a vulnerable area and is treated by the neuro-oncology team at Máxima,' says Van Santen. 'The quality of life after surgery is determined by the degree of damage to the hypothalamus. This can be damaged by the tumor, but also by the surgery.' Together with Máxima colleagues Dr. Marc van de Wetering and Prof. Eelco Hoving, Van Santen is investigating how this damage can be prevented or limited as much as possible. For example, in the near future we will be starting a new technique called intra-operative MRI that will allow us to look at the tumor, or what is left of it, during surgery. We are also working on developing an organoid model, a mini-organ grown from a child's cells, on which we can test different drugs. If we can treat a craniopharyngeoma with drugs, we would need to perform less frequent and less invasive surgery. Radiation would also be needed less often. With this we hope to prevent damage to the hypothalamus. We are also introducing new treatments for hypothalamic obesity, a side effect caused by the hormone changes.'

Prevention

To provide children with a hypothalamic syndrome with optimal guidance, Van Santen has set up a multidisciplinary team. In it, the pediatric endocrinologist, psychologist, child psychiatrist, dietician, physiotherapist, neurologist, rehabilitation specialist and oncologist work closely together. 'As a team, we are building expertise regarding this rare disorder. We are also gaining more insight into the symptoms. These differ per patient,' she says. If alarm signals are better recognized we can use an MRI to detect small tumors in the pituitary gland earlier and they are easy to treat. Once the hypothalamus is damaged, however, you have a problem. A damaged hypothalamus leads to all kinds of consequences of a disturbed hormone balance, for example (morbid) obesity, behavioral problems and diabetes insipidus without an adequate sense of thirst which is very difficult to treat.

European collaboration

'My goal is to offer patients with hypothalamic dysfunction better treatment in the future,' says van Santen. 'The condition is rare, so research and increasing knowledge requires collaboration. As with many rare disorders, it helps to set up international studies. I hope to set up a European consortium so that we can jointly investigate which interventions can help these patients.' To encourage collaboration, we are organizing the European Science symposium on hypothalamic dysfunction from the European Society for Endocrinology (ESPE) at Máxima on October 7 and 8. 'During these two days we will speak with several renowned international experts about scientific developments and future cooperation.'

More awareness is also needed about the hypothalamic syndrome so that it can be recognized earlier. That is why Van Santen, together with fellow physician and professor Dr. Hermann Müller (Oldenburg, Germany), is developing a tool to recognize patients earlier and then classify them in the same way. 'The picture differs per patient, but I am convinced that if we recognize certain patterns better and earlier we can start to mean more for these children.'