**The current and future care management of the congenital central hypoventilation syndrome in Japan**

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In this session, I would like to discuss mainly about care management for CCHS in Japan.

**【Social Insurance System】**

We have unique social health care insurance systems, and have to register one of them. These insurance systems involve illness, injury, childbirth, and death for the insured person as well as his or her dependents, though we have to pay 30% of the total medical expenditure generally. In addition, we can visit a medical institution which is clinic or also hospital at any time.　However, recently, medical costs are unnecessary for children up to the 12 or 15 years old depends on their living area. They are provided by local and/or national government.

**【Management and Treatment by Pediatrician】**

Pediatrician and also neonatologist look after most of CCHS patients, and also lead care for CCHS patients by themselves. Most of them are working in reference hospital which is acceptable for hospitalization, and general practitioner is a few.

**【Management and Treatment Strategy】**

Because the management methods of CCHS are divergent and not integrated, they are performed depending on the condition of CCHS patient to evaluate arterial blood gas analysis, echocardiography, spirograph, electroencephalogram and genetic analysis and to manage for ventilator setting, facial interface, tracheostomy and cardiac pacing. Of course, patients are admitted to the intensive care unit if necessary.

The plan of at-home care will be indicated by medical staffs but it will be decided by both medical staffs and also the patients and their families. Although technical support and management for ventilator are provided by distributers, the patients or their families are instructed to be able to operate the setting. Home monitoring with pulse oximeter is used generally, however the cost for monitoring is not included in insurance systems.

**【Management for Teenager and Adult Patients】**

Thus far, most patients of teenager or adult are looked after by pediatricians who have continued to lead care. In future, physician for adult patients might provide medical management for the CCHS patients.

**【Future Direction】**

Despite progress with regard to respiratory care management, neurodevelopmental disability is often seen. Because the hypoxemia will be considered to affect central nervous system, further investigation for neurodevelopmental condition of CCHS patients is strongly demanded. By integrating for appropriate medical examination and treatment, we are expected to suppress the influence on neurodevelopment to the minimum.