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RASTELLI-SENNING REPAIR FOR RARE CONGENITAL HEART DEFECT

Heart surgeons from KK Women's and Children's Hospital successfully perform complex Rastelli-Senning procedure on teen to repair rare variant of congenitally corrected transposition of the great arteries.

A cardiothoracic surgery team at KK Women's and Children's Hospital (KKH) has successfully performed a complex Rastelli-Senning surgical procedure to repair the heart of a 16-year-old patient with a rare congenital abnormality.

Born with a rare variant of congenitally corrected transposition of the great arteries (ccTGA), the patient's cardiac ventricles were reversed during fetal development – resulting in each ventricle being connected to the incorrect atrium and great artery. The condition was compounded by a large hole in the patient's ventricular septum and a stenosed pulmonary valve.

Due to the abnormal cardiac blood flow pathway and lack of blood permitted to the lungs, the patient was frequently cyanotic due to low levels of oxygen in the body. While able to attend school, the patient experienced poor effort tolerance and was unable to participate in any activity requiring physical exertion.

"Carefully considering the patient's very impaired quality of life and the almost-certain prospect of heart failure in their 40s, our care team of radiology, cardiology and cardiothoracic surgery specialists decided to proceed with the Rastelli-Senning procedure to surgically repair the patient's heart and correct the cardiac blood flow," shared Dr Loh Yee Jim, Consultant, Cardiothoracic Surgery Service, KKH, who led the surgical procedure.

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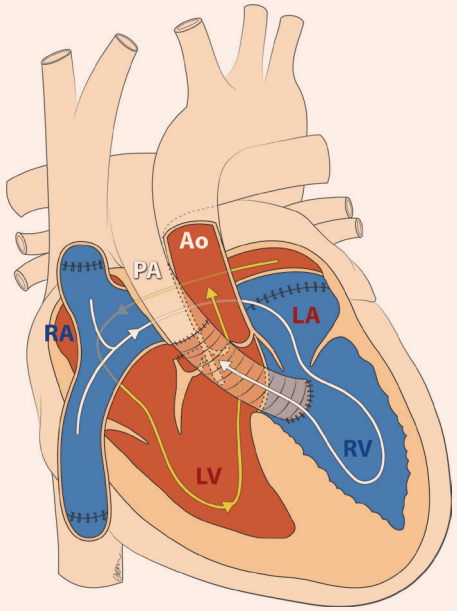

Heart with ccTGA, ventricular septal defect and pulmonary stenosis after Rastelli-Senning repair.

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RASTELLI-SENNING PROCEDURE TO REPAIR CCTGA AT KKH

Following a thorough diagnostic evaluation to determine the patient's suitability for the highly complex surgery, a cardiothoracic surgical team performed the multi-step Rastelli-Senning procedure to reroute the patient's blood flow through the correct ventricles and great arteries.

First, the Senning procedure was carried out to reroute cardiac blood flow from each atrium to the correct ventricle. This involved the creation of a baffle within the atria to redirect the deoxygenated caval blood to the tricuspid valve and the oxygenated pulmonary venous blood to the mitral valve. Then, the Rastelli procedure was performed. This involved closing the sizeable hole in the patient's ventricular septum at an angle, to direct blood flow from the left ventricle to the aorta; and the insertion of a valve conduit into the right ventricle to direct blood flow to the pulmonary artery.

Post-surgery, the patient was temporarily placed on extra-corporeal membrane oxygenation (ECMO) support to allow time for the left ventricle to accustom itself to the increased pressure and blood flow. The patient's heart soon stabilised and was able to function without external support.

In the weeks following the operation, the patient underwent physiotherapy, respiratory therapy and dietetic advice to facilitate full recovery. Currently, the patient is asymptomatic and able to participate in recreational activities requiring moderate levels of effort.

"The successful Rastelli-Senning procedure allows patients with ccTGA to have a much better quality of life, with very much improved exercise tolerance. Most importantly, as the patient's blood now flows through the correct chambers of the heart, this greatly reduces the likelihood of heart failure and the need for a heart transplant later, during their prime years," said Dr Loh.

"Due to the complexity of the condition, patients with ccTGA require life-long follow-up. Our patient will be monitored by the KKH care team until adulthood. We will then work closely with the adult congenital team at the National Heart Centre Singapore to ensure our patient's seamless transition into their care."

THE SENNING AND RASTELLI PROCEDURES

The Senning procedure rerouted cardiac blood flow from each atrium to the correct ventricle. This involved the creation of a baffle within the atria to redirect deoxygenated caval blood to the tricuspid valve and oxygenated pulmonary venous blood to the mitral valve.

The Rastelli procedure was performed to close the hole in the patient's ventricular septum, directing blood flow from the left ventricle to the aorta. A valve conduit was inserted into the right ventricle to direct blood flow to the pulmonary artery.

CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

A rare congenital heart abnormality, ccTGA occurs in 0.5 percent of patients diagnosed with congenital heart disease. The condition is characterised by the reversal of the ventricles during fetal development, resulting in each ventricle being connected to the incorrect atrium and incorrect great artery.

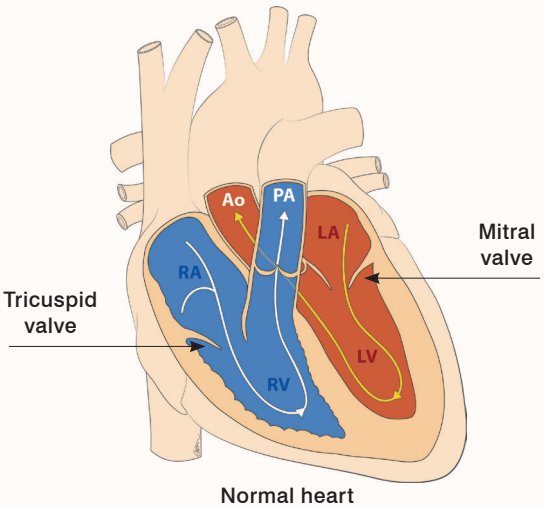
This complex malformation places great strain on the patient's heart – particularly on the smaller right ventricle, which is forced to bear a greater blood pressure and volume than it was genetically meant to. This can lead to further health problems, such as irregular heart rhythm, valvular regurgitation, impaired heart function and heart failure.

Several associated problems can also co-exist in patients with ccTGA, potentially further complicating their quality of life and the type of intervention that is required. These can include:

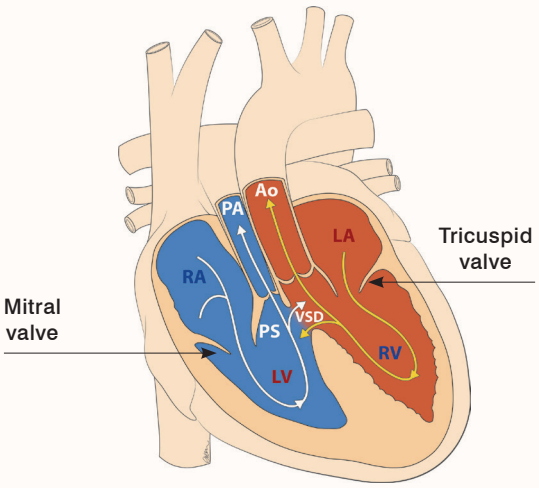
- Ventricular septal defect – a hole between the ventricles of the heart
- Pulmonary stenosis – obstruction of blood flow from the right ventricle to the pulmonary artery
- Leakage of the tricuspid valve
- Obstructed cardiac electrical conduction system

Medical literature reports that 67 percent of patients with ccTGA and associated heart defects develop heart failure by the age of 45, eventually requiring a heart transplant. However, with appropriate surgical intervention, over 90 percent of patients with ccTGA can have a 20-year survival.

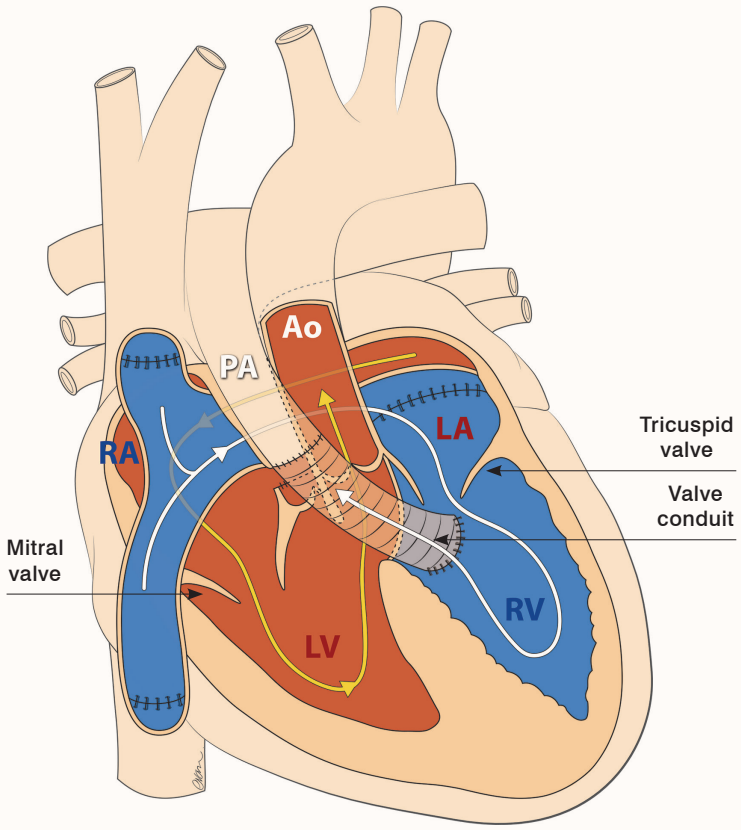
ILLUSTRATION OF THE RASTELLI-SENNING PROCEDURE



Normal heart



Heart with ccTGA, ventricular septal defect and pulmonary stenosis.



Heart with ccTGA, ventricular septal defect and pulmonary stenosis after Rastelli-Senning repair.

LEGEND

RA	Right atrium	Ao	Aorta
RV	Right ventricle	PA	Pulmonary artery
LA	Left atrium	PS	Pulmonary stenosis
LV	Left ventricle	VSD	Ventricular septal defect

CRITICAL CARE FOR CONGENITAL HEART ABNORMALITIES

Congenital heart abnormalities are the most common structural anomalies among babies born in Singapore, occurring in nine per 1,000 live births. At KKH, babies and children requiring critical care for heart and lung conditions are managed by comprehensive multidisciplinary teams comprising radiology, cardiology, cardiothoracic surgery, paediatric and neonatal intensive care, anaesthesia and allied health specialists.

The KKH team performs the largest number of complex congenital heart surgeries for babies, children and adults in Singapore, and also provides critical cardiac and thoracic care for many patients in Singapore and the region. Despite this, the hospital maintains the lowest mortality rate for paediatric cardiac surgery in Singapore, with a core success rate of 97-98 percent over the last five years.

The hospital's ECMO programme also provides life-sustaining support to young patients requiring emergency care for high-risk heart and lung conditions. Singapore's first mobile ECMO service for infants and children was recently created by KKH to assist patients urgently requiring ECMO support at other locations before being brought to the KKH intensive care unit for ongoing care.

To advance cardiac care for the region, the KKH team also provides training and medical expertise to assist the establishment of congenital cardiac programmes in countries such as Myanmar and Vietnam.

"DOCTOR, IS MY BABY NORMAL?"

Prenatal risk assessment for Down syndrome

Professor George SH Yeo, Head and Senior Consultant, Department of Maternal Fetal Medicine, KK Women's and Children's Hospital

One of the most common questions pregnant women ask their obstetrician is, "Doctor, is my baby normal?" While most pregnancies proceed without incident, women of any age can have a small risk of their baby being affected by congenital abnormalities. Generally, the incidence of congenital abnormalities can be categorised into: structural (2%), syndromic (0.5%) and chromosomal (1%). The most common chromosomal abnormality found at birth is Trisomy 21, commonly known as Down syndrome.

DOWN SYNDROME

In Singapore, Down syndrome occurs in one of every 500 births. Almost 50 percent of children with Down syndrome have structural or physical disabilities, some of which may be surgically correctable. The majority have low cognitive ability and mild to moderate developmental disabilities, and a smaller number have severe to profound mental disability. The estimated global incidence of Down syndrome ranges from one in 700 to one in 1,000 live births. The one-year survival rate of children with Down syndrome is 85 percent, and 40 percent are expected to live longer than 50 years.

Advancements in screening technologies and management of early pregnancy in recent years have made it possible to offer accurate screening for Down syndrome to pregnant women of all ages. The risk of a fetus having Down syndrome can be assessed by several types of tests. These include non-invasive screening tests such as the first trimester screening test (FTS) and non-invasive prenatal test (NIPT)*, and invasive diagnostic tests such as amniocentesis and chorionic villus sampling (CVS).

FIRST TRIMESTER SCREENING

First trimester screening test (FTS) is a combined screening test that assesses the risk of a fetus having Down syndrome, other chromosomal disorders and fetal structural defects. Performed between 11 and 14 weeks of gestation, FTS calculates a composite chromosomal risk score based on maternal age, measurement of fetal nuchal translucency (NT) and the fetal nasal bone, and the levels of free beta-human chorionic gonadotrophin (beta-hCG) and pregnancy-associated plasma protein A (PAPP-A) in the maternal blood.

The FTS combined test is globally recognised as the gold standard for Down syndrome risk assessment during pregnancy. At KK Women's and Children's Hospital (KKH), FTS is conducted for over 6,000 women a year, with a detection rate of above 95 percent for Down syndrome over the last 10 years.

Maternal age

Studies show that a woman's chances of having a child with Down syndrome increase with age, due to a greater risk of improper chromosome division in the egg. By age 35, a woman's risk of delivering a child with Down syndrome is about one in 350. By age 40, the risk is about one in 100, and by age 45, the risk is about one in 30.

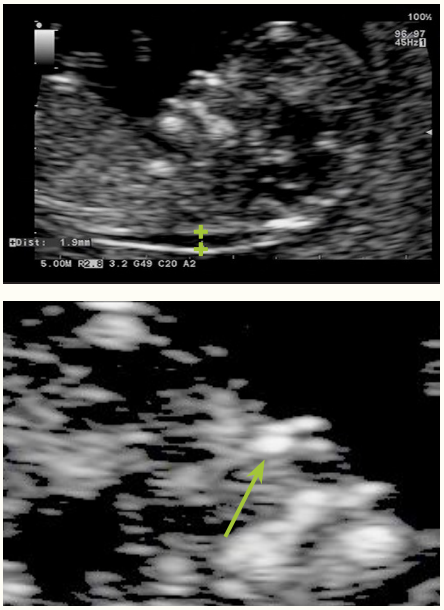
Fetal nuchal translucency

Measured by ultrasound, the thickness of the skin behind the fetal neck – also known as nuchal translucency or NT – is a marker in the estimation of the risk of Down syndrome. Seventy-five percent of fetuses with Down syndrome are noted to exhibit increased NT. However, increased NT is not a definite diagnosis of Down syndrome, as five percent of normal fetuses are also noted to exhibit this characteristic.

In addition to Down syndrome, increased NT may also be an indicator of other fetal chromosomal abnormalities and is associated with a number of structural abnormalities and genetic syndromes.

Free beta-hCG and PAPP-A levels in maternal blood

Free beta-hCG and PAPP-A are maternal serum biochemical markers for Down syndrome. During FTS, these are measured via a maternal blood test. Typically, in pregnancies affected by Down syndrome, the level of maternal beta-hCG is increased and the level of PAPP-A is reduced as compared to a normal pregnancy with a similar gestation period.



The nuchal translucency (top) and nasal bone (bottom) of a normal fetus, both indicated in green.

The integrated approach offered by FTS enables physicians to detect structural abnormalities, as well as a range of other chromosomal abnormalities in the fetus. As FTS is a screening test, should it indicate a high risk result – a risk of one in 300 or higher – a diagnostic test such as amniocentesis or CVS will be offered for exclusion or confirmation of the condition.

NON-INVASIVE PRENATAL TEST

The non-invasive prenatal test (NIPT) is a recently-developed screening test for Down syndrome which assesses fetal DNA in the maternal bloodstream. This blood test is typically performed between 10 and 22 weeks of gestation.

The NIPT has a sensitivity and specificity above 99 percent in the detection of Down syndrome.

While this makes it a very effective screening test for Down syndrome, it is important to note that the NIPT is unable to detect any structural anomalies and many chromosomal anomalies in the fetus. Like the FTS, the NIPT is also unable to provide a definitive diagnosis of Down syndrome. Should the NIPT indicate a high risk result, a diagnostic test such as amniocentesis or CVS will be offered for exclusion or confirmation of the condition.

AMNIOCENTESIS

Amniocentesis is a diagnostic test offered to women who are at higher risk of having a baby with Down syndrome as determined by a screening test. The test is performed when the fetus is beyond 16 weeks of gestation, and involves a procedure in which amniotic fluid is drawn with a needle from the uterus to test the cells.

Due to its invasive nature, amniocentesis generally carries a procedure-related risk of miscarriage, ranging from one in 300 to one in 50. Test results can reliably rule out or provide a diagnosis of Down syndrome. Clinical audits at KKH indicate the hospital's amniocentesis-related risk of miscarriage to be one in 300.

CHORIONIC VILLUS SAMPLING

Chorionic villus sampling (CVS) is also a diagnostic test offered to women who are at higher risk of having a baby with Down syndrome. The test is commonly performed after 11 weeks of gestation, and involves removing a sample of chorionic villi tissue from the placenta for testing. CVS generally carries a procedure-related risk of miscarriage similar to amniocentesis. Test results can reliably rule out or provide a diagnosis of Down syndrome.

SCREENING IS KEY TO EARLY DETECTION OF DOWN SYNDROME

While undergoing screening for Down syndrome is not compulsory, it is strongly recommended, as screening tests are non-invasive and accurate ways of assessing the risk of Down syndrome in a fetus. Additionally, the individualised risk score afforded by FTS allows for more precise prenatal counselling, empowering women to make informed decisions regarding the subsequent management of their pregnancy. Since 1999, KKH has had a dedicated team of prenatal screening counsellors, who provide counselling to pregnant women regarding screening and diagnostic testing for Down syndrome.

It is important to note that the vast majority of screen-positive test results are false-positive, i.e. even when a screening test indicates a high risk result, e.g. one in 50, the fetus may not necessarily have Down syndrome. A diagnostic test is required to either exclude or confirm the diagnosis of Down syndrome.

Screening fulfills the crucial purpose of identifying women whose risk of having a baby with Down syndrome is sufficiently high to necessitate an invasive diagnostic test. This is important in order to minimise the need for invasive testing for most pregnancies. At KKH, more than nine out of 10 patients are assessed as lower risk for Down syndrome by FTS and therefore do not require invasive testing. Even amongst pregnant women above the age of 35, four out of five are assessed as low risk for Down Syndrome by FTS.

Early detection of Down syndrome or other congenital abnormalities provides the option of preparing couples psychologically for selective procreation as well as the birth and care requirements of a child with special needs. The diagnosis also allows physicians to initiate early prenatal counselling and support to couples, and to tailor their management of the pregnancy to provide optimal care to the mother and baby well in advance of labour and delivery.

A pioneer in the fields of obstetric care and fetal medicine in Singapore, Professor George SH Yeo is instrumental in establishing many clinical protocols and guidelines for prenatal diagnostic services and prenatal screening over the last 25 years in Singapore. In addition to his roles as Chief of Obstetrics, Head of the Obstetric Ultrasound and Prenatal Diagnosis Unit and Director, Antenatal Diagnostic Centre, KKH, Prof Yeo also currently serves as Adjunct Professor, Duke-NUS Graduate Medical School, and Clinical Associate Professor, Yong Loo Lin School of Medicine.

A strong advocate of clinical research, Prof Yeo has close to 160 publications in indexed journals and is Director of Research, SingHealth-Duke NUS Academic Clinical Programme for Obstetrics & Gynaecology. He is also Chairman, Advisory Committee, National Birth Defects Registry, Ministry of Health, Singapore.

*The term 'non-invasive prenatal test' (NIPT) in this article refers to the fetal DNA screening test for Down syndrome.

A UNIQUE WAY OF LEARNING

Effective learning strategies for children with developmental needs

MaryAnne Ho, Senior Psychologist, Department of Child Development, KK Women's and Children's Hospital

Developmental needs encompass a diverse group of severe chronic conditions that are caused by mental and physical impairment. Children with developmental needs may experience difficulties in several areas, including mobility, self-help, independent living, language and learning. These difficulties usually persist throughout the child's life[1].

Early detection and intervention have been shown to improve the well-being and coping mechanisms of children with developmental needs. In particular, children with mild developmental needs and non-impaired levels of cognitive functioning are generally able to participate in mainstream education with appropriate help. This includes children with attention deficit hyperactivity disorder (ADHD), autism spectrum disorder (ASD) and language impairment.

STRUCTURED LEARNING HELPS CHILDREN WITH ADHD

Children with ADHD exhibit behaviours such as inattention, hyperactivity and impulsivity of a severity and frequency greater than that expected of their developmental age. According to the Diagnostic and Statistical Manual of Mental Disorders-IV (DSM-IV), ADHD is diagnosed when a child exhibits a pattern of such behaviours in two different settings for six months or more[2].

While children with ADHD are frequently of average intelligence, these associated behavioural patterns often result in them experiencing learning difficulties.

Maintaining a learning environment that is structured, with rules, schedules and expectations aids effective learning for a child with ADHD. Some strategies include:

- Seating the child in a location and arrangement that minimises sensory and social distractions while providing the educator optimal opportunity to monitor the child's behaviour and redirect their attention.
- Implementing positive reinforcement to encourage appropriate behaviours[3].
- Applying consequences consistently and in a timely manner when the child exhibits inappropriate behaviour e.g. withholding a privilege if the child does not complete their homework.
- Teaching the child self-monitoring strategies such as 'Stop, Think, Do'[4] (Figure 1).
- Teaching the child metacognitive strategies to aid learning and comprehension. This can include leading the child in predicting questions to be answered during reading, helping the child to define unfamiliar terms and putting the main ideas into brackets.

VISUAL INSTRUCTION HELPS CHILDREN WITH ASD

Based on the DSM-IV, ASD is characterised by developmental delays or abnormal functioning in one or more domains of social interaction and communication before the age of three years.

Children with ASD often exhibit restricted, repetitive and stereotyped patterns in their behaviour, interests and activities. These include an insistence on following routines or rituals, repetitive motor mannerisms, and an encompassing preoccupation with specific patterns of interest, resulting in the exclusion of others.

In addition, children with ASD experience impairment in maintaining reciprocal communication. This includes difficulty with using body language such as gestures, facial expressions and body postures when communicating with others. As a result, children with ASD may experience difficulty with responding socially or emotionally to social overtures, engaging in and sustaining reciprocal conversations and developing age-appropriate peer relationships.

When facilitating learning for a child with ASD, it is important to take into account their sensory needs, short attention span and lack of planning and organisational abilities. Arrangements to aid learning can include:

- Seating the child in a dimmer and quieter room if the child is hypersensitive to light and sound.
- Using visual aids and instruction methods (Figures 1 and 2).
- Developing an individualised work system and daily schedule for the child.
- Incorporating a topic of specific interest to the child, which can help to differentiate the curriculum and provide motivation for learning.
- The use of social stories[5] to explain to the child how others perceive a social situation and to suggest alternative ways of managing the situation.

Figure 1. Illustration of the 'Stop, Think, Do' approach, which visually prompts the child to pause before acting and think carefully of the possible options and alternatives before choosing the most appropriate action.

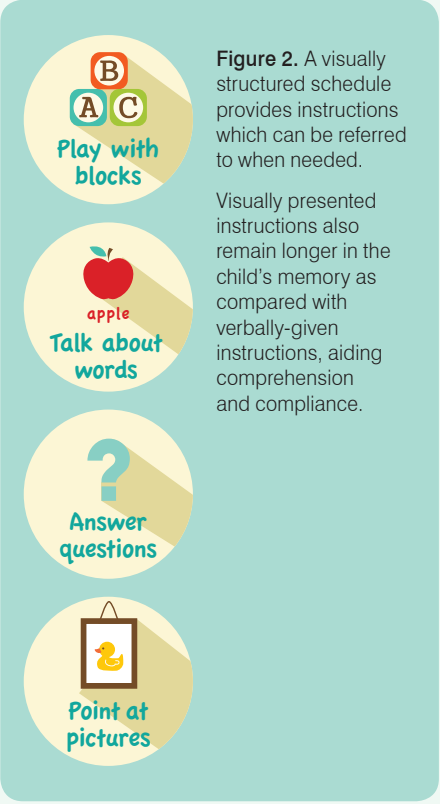


Figure 2. A visually structured schedule provides instructions which can be referred to when needed.

Visually presented instructions also remain longer in the child's memory as compared with verbally-given instructions, aiding comprehension and compliance.

FUNCTIONAL PRACTICE HELPS CHILDREN WITH LANGUAGE IMPAIRMENT

Language impairment affects about seven percent of children[6], and is characterised by delayed or disordered language for no apparent reason, despite the child possessing adequate cognitive abilities[7].

Due to its developmental nature, language impairment may manifest as delayed speech during toddlerhood, difficulty with understanding grammar during preschool years and limited fluency and understanding of language during school years. Some children also experience difficulties with comprehending spoken language that is abstract or complex.

As language and literacy are closely linked, children with language impairment frequently experience difficulty with learning written language[8]. Strategies to aid effective learning for a child with language impairment include:

- Speaking in shorter sentences and writing out sentences if the child has difficulty comprehending long sentences.
- Explicitly teaching the child vocabulary related to specific topics; this is especially useful for abstract topics.
- Providing the child opportunities to practice various grammatical constructs or narrative language.
- Checking whether the child understands a "wh-" question by asking them to restate the question in their own words.
- Explicitly teaching the child non-literal uses of language, such as idioms, and metalinguistic skills such as phonemic awareness[3].

Contemporary approaches to language impairment weave intervention into natural episodes of communication. Such intervention builds on the child's utterances to develop their social use of language within a functional context.

Parents are encouraged to be involved in their child's intervention and provide opportunities within the child's daily routines to practice their language skills. At KK Women's and Children's Hospital, speech language therapists provide extensive training to assist parents with incorporating intervention into the child's environment.

POSITIVE, ENLIGHTENING EXPERIENCES AID EFFECTIVE LEARNING

Although children with developmental needs have unique learning needs, educational methods which take into account their strengths and address their weaknesses can help educators to create a positive and enlightening learning experience for them.

As with all children, it is important to acknowledge and recognise each child's strengths and to provide them with the opportunities to build on these. This will in turn enhance their self-esteem and help them to meet their full potential.

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MaryAnne Ho graduated from Murdoch University in Perth, Western Australia, and completed a Master of Applied Psychology at Nanyang Technological University. She has a special interest in autism spectrum disorder and learning difficulties.

MANAGEMENT OF HEARING IMPAIRMENT IN CHILDREN

Associate Professor Henry Tan, Head and Senior Consultant, Department of Otolaryngology, KK Women's and Children's Hospital

Hearing impairment is a common disability, with one in 1,000 babies in Singapore experiencing significant hearing impairment at birth. This places them at risk for delayed speech, language, intellectual, social and emotional development. However, timely intervention can effectively maximise the child's communication potential if instituted early in infancy.

In recent years, the implementation of Universal Newborn Hearing Screening (UNHS) in hospitals worldwide has facilitated early detection and intervention for many babies with hearing impairment. The UNHS is a hearing assessment programme which screens newborns for hearing impairment within the first few days following birth.

COMMON TYPES OF HEARING IMPAIRMENT IN CHILDREN

There are various types of hearing impairment, all of which can occur unilaterally or bilaterally. Two of the most common types of hearing impairment are sensorineural hearing loss (SNHL) and conductive hearing loss.

SNHL is the most common hearing impairment diagnosed via UNHS, with a global prevalence of 0.5 to one per 1,000 live births, increasing to 1.5 to two per 1,000 children by six years. SNHL is due to damage to the cochlea or to the nerve pathways from the inner ear to the brain. These can be due to one or more of the following causes:

- Prenatal genetic causes – There are approximately 70 syndromes associated with SNHL, of which the Down and Goldenhar syndromes are most commonly seen in Singapore.
- Prenatal non-genetic causes – These include maternal infections, such as the TORCHES (toxoplasmosis, other, rubella, cytomegalovirus and herpes simplex) group of diseases, alcohol and drugs such as aminoglycosides and thalidomide.
- Perinatal causes – These include prematurity, low birth weight of less than 2.25 kilograms, anoxia, hypoxia and hyperbilirubinemia.
- Postnatal causes – These include sensorineural, infective, idiopathic, viral, vascular and noise-induced issues, ototoxicity, myelofibrosis with myeloid metaplasia, Meniere's disease, temporal bone fracture, perilymph leak and acoustic neuroma.

Unilateral SNHL may be congenital, or due to causes such as mumps or head injury. Children with unilateral SNHL usually do not have any notable hearing or speech impairment. The hearing impaired ear often goes unnoticed until the hearing in the normal ear is affected.

Conductive hearing loss occurs when sound is not conducted efficiently through the outer ear canal to the eardrum and the ossicles of the middle ear. The condition can occur due to an infection of the middle ear secondary to eustachian tube dysfunction, such as acute or chronic otitis media with effusion, or chronic suppurative otitis media. Other causes include trauma and otosclerosis.

Congenital conductive hearing loss may be due to meatal stenosis or atresia with or without microtia, ossicular chain abnormalities, congenital cholesteatoma or osteogenesis imperfecta.

ASSESSMENT FOR HEARING IMPAIRMENT IN CHILDREN

Clinical history taking is important when assessing a child for hearing impairment. The physician should enquire about cardinal symptoms of the ear, nose and throat such as hearing loss, tinnitus, vertigo, ear discharge and associated nose or throat complaints. The physician should also enquire about prenatal infection (TORCHES), term or premature pregnancy, birth weight, neonatal jaundice, a family history of hearing loss, problems with the child's central nervous system and other medical history.

As all body systems have been associated with SNHL, symptoms can occur in isolation or as part of a syndrome. Thus physical examination should extend beyond the ears, nose and throat to include other body systems – particularly the eyes and neurological system.

Signs which may indicate SNHL include:

- **Ear:** auricular appendages or malformation
- **Eye:** coloboma, cataract, retinitis pigmentosa
- **Neck:** goiter
- **Musculoskeletal:** fusion of the cervical vertebrae
- **Integumentary system:** white forelock, hypopigmentation
- **Neurological system:** ataxia, mental disability

There are several types of hearing tests that can be done at any age. These include tympanometry, otoacoustic emission, automated auditory brainstem response and steady-state evoked potential (SSEP).

The tuning fork test can further help to differentiate conductive hearing loss from SNHL.

However, this test is only applicable to children, six years and above, and test results must be judged with care, as children may not always be able to cooperate to give a reliable result. Other hearing tests include the distraction test, which can be used for children older than six months; conditioning audiometry, which can be carried out from two to five years and pure tone audiogram, which can be carried out from four years.

Imaging studies, such as computerised tomography or magnetic resonance imaging, are useful to investigate SNHL and conductive hearing loss. Imaging allows physicians to assess the facial nerves, middle and inner ears, such as the auditory ossicles, otic capsules and internal auditory canal. Such studies are useful to the physician when counselling patients about their condition, as well as for the management of mixed and profound hearing loss.

A range of laboratory tests can be carried out for the assessment of hearing impairment. These include full blood count, renal panel, thyroid function, fluorescent treponemal antibody absorption (FTA-ABS), urinalysis, intrauterine infection, metabolic work-up, autoimmune work-up and the work-up of the vestibular portion of the inner ear. The use of these tests should be based on the physician's index of suspicion.

MANAGEMENT OF HEARING IMPAIRMENT IN CHILDREN

When a newborn is diagnosed with a hearing impairment, early intervention is crucial to maximise the child's communication potential. A range of interventions can be implemented, such as hearing amplification, auditory verbal therapy, natural auditory oral therapy and cochlear implants.

Additionally, continued professional guidance and help should be extended to the child and their family to assist with the child's development and education. Relevant healthcare providers who can provide integrated support to the child and their family consist of the otologist, family physician, paediatrician, teacher for the hearing impaired, educational psychologist, speech pathologist and community partners.

Hearing aid

Most children with hearing impairment will benefit from a hearing aid, which should be fitted early. There are several types of hearing aids, such as post-aural, in-ear or radio aids. Regardless of the choice of hearing aid, binaural use is usually recommended. Regular adjustment and counselling is crucial to encourage the child's continued use of the device.

Cochlear implant

Cochlear implantation provides direct electrical stimulation to the auditory nerve in the inner ear, and can be beneficial for hearing impaired children in whom the use of hearing aids is not possible. Early cochlear implantation in children has been shown to result in superior speech and language outcomes. Since the establishment of the cochlear implant programme at KK Women's and Children's Hospital (KKH) in 2005, the mean age of cochlear implantation at the hospital has reduced from nine to three years.

Education

Access to education and rehabilitative services is critical for hearing impaired children to maximise their speech, language and academic outcomes. Supportive teachers, continual assistance from a teacher for the hearing impaired and preferential seating are important to facilitate optimal learning and development for the hearing impaired child. The educational setting must also provide an auditory environment with a range of communication modes – including oral speech, hand signs and body language – to enable the hearing impaired child to fully utilise their remnant hearing.

COMMON SYMPTOMS OF HEARING IMPAIRMENT IN BABIES AND CHILDREN

Hearing impairment in babies or children may present in one or more of the following ways:

- Quietness
- Not being startled by loud sounds
- Delay in speech development
- Speech defects or changes
- Non-responsiveness to verbal instruction
- Misbehaviour
- Academic difficulties

Generally, a child should be able to speak two to three words by 12 months; 20 words by 18 months; be able to string words together by 21 to 24 months and have a vocabulary of about 250 words by two to five years.

The child should be referred to seek tertiary assessment should they be unable to speak by 18 months or be unable to form sentences by 30 months.

A firm advocate of life-long learning, Associate Professor Henry Tan is active in medical education and research. He currently serves as Associate Professor, Duke-NUS Graduate Medical School as well as Clinical Associate Professor, Yong Loo Lin School of Medicine. Assoc Prof Tan has also published widely, with more than 60 publications to date. Assoc Prof Tan is the leading cochlear implant surgeon for children with hearing loss at KKH.

HELPING NEWBORNS TAKE THEIR FIRST BREATH

In pursuit of excellence in neonatal care, KK Women's and Children's Hospital (KKH) has been accredited by the National Resuscitation Council of Singapore (NRCS) as a neonatal resuscitation training centre for healthcare professionals involved in the delivery and care of a newborn.

More than 90 percent of infants have an uneventful birth. However, about one in ten newborns experience difficulty with the transition to extra-uterine life, and require some assistance to begin breathing.

Perinatal asphyxia and extreme prematurity are two complications of pregnancy that most frequently necessitate complex resuscitation by skilled personnel. Additionally, many infants with low birth weight require some stabilisation and assistance at delivery.

Neonatal resuscitation aims to prevent morbidity and mortality associated with injury of the brain, heart and kidney, and to re-establish adequate spontaneous respiration and cardiac output in a newborn.

INTEGRATED RESPONSE FOR NEONATAL RESUSCITATION

Following accreditation by the NRCS, a multidisciplinary team at KKH has begun facilitating the Singapore Newborn Resuscitation Course (SNRC) for the hospital's 500-strong team of physicians and nurses involved in the care and resuscitation of newborns.

"Preparedness is the first and most important step in delivering effective neonatal resuscitation," shares Dr Quek Bin Huey, Senior Consultant, Department of Neonatology, KKH, who led the accreditation effort. "At KKH, all medical personnel who provide care for infants at birth and during the neonatal period receive regular and standardised training in neonatal resuscitation. This enables the attending medical team to provide an immediate and integrated response should the need arise. The SNRC is a welcome boost to enhance the rigour of our existing training programme."

Completion of the SNRC is recommended as part of best practices for all healthcare professionals involved in neonatal care. The education programme involves interactive online lectures and training modules, as well as practical demonstrations and simulation training in a variety of clinical scenarios. Detailed post-simulation evaluations are conducted to provide participants real-time feedback akin to a live bedside situation.

"Successful neonatal resuscitation requires a concerted effort, and involves not only clinical knowledge, but also key behavioural skills, communication and teamwork," says Dr Quek. "The dissemination of standardised training in neonatal resuscitation through the SNRC can help to ensure all healthcare providers involved in neonatal care acquire and maintain the skills necessary for effective neonatal resuscitation. This is a crucial educational step in improving patient outcomes."

KKH is one of only two training centres in Singapore which have been accredited by the NRCS to facilitate training in neonatal resuscitation. The hospital also plans to extend the accredited programme to other healthcare providers.

Paediatric Surgery Forum

GP Forum for Paediatric Health 2014

Date : 22 November 2014 (Saturday)
Time : 1.00pm to 5.00pm
Fee : $10 per pax (Includes lunch, tea and parking)
Venue : KKH Auditorium, Training Centre, Level 1, Women's Tower

For full details of KKH forums, scan this code with your smartphone now.

For more details, please call **+65 6394-8746** (Monday to Friday, 8.30am to 5.30pm) or log on to **www.kkh.com.sg/events**

EDUCATING FOR A HEALTHIER FUTURE FOR SINGAPORE

For her distinguished and outstanding contributions to the development of paediatric medicine in Singapore, Professor Chay Oh Moh was awarded the National Outstanding Clinician Mentor Award 2014 under the National Medical Excellence Awards, in August 2014.

As a child, Professor Chay Oh Moh observed a cousin suffering repeated acute asthma attacks, and wondered why no one had taught her family how to manage and prevent the attacks.

"The family's first aid box contained home remedies for fever, colds, cough, pain and diarrhoea – but they lacked the knowledge to manage their child's acute asthma," she said.

Years later, while undergoing training in paediatric medicine, Prof Chay again noticed a need – this time for respiratory medicine specialists in public healthcare.

"I decided to focus my efforts on enhancing care for children by advancing clinical practice and public awareness of respiratory medicine in Singapore."

Believing in the power of education to help her reach her goal, Prof Chay set to the task of raising a successive generation of healthcare professionals who would be dedicated not only to holistic and compassionate care for patients, but also to enabling better health for future generations.

In addition to her clinical duties, she undertook the development and mentorship of countless medical students and residents in the areas of paediatrics and paediatric respiratory medicine. She also became the first Academic Chair of the SingHealth-Duke NUS Paediatric Academic Clinical Program (2012-2013), a SingHealth-wide framework for the advancement of education, research and clinical care in paediatric medicine.

Prof Chay remains hard at work improving care and the quality of life for patients with respiratory problems, currently serving as Deputy Program Director for the Singapore National Asthma Program (SNAP). Together with her team, she advocates for collaborations between healthcare providers, patients, their families and community partners to facilitate the home management of asthma.

> "Our mandate is to equip the next generation to succeed and surpass us in knowledge, clinical skills, collaboration and most importantly, compassion for the sick and the wounded.
>
> Together with countless other doctors, nurses, allied health professionals and researchers, we continue to teach, train and mentor all who play a part in the health of our nation."
>
> **Professor Chay Oh Moh**

"An individual, no matter how committed, can only help a limited number of patients in their lifetime," said Prof Chay. "In order to drive effective and sustainable care for patients now and in the future, a multiplier effect is necessary through the integrated education and training of healthcare providers, community partners, patients and caregivers."

"Our mandate is to equip the next generation to succeed and surpass us in knowledge, clinical skills, collaboration and most importantly, compassion for the sick and the wounded. Together with countless other doctors, nurses, allied health professionals and researchers, we continue to teach, train and mentor all who play a part in the health of our nation."

Professor Chay Oh Moh continues to contribute greatly to the advancement of paediatric medicine and education in Singapore as Campus Director, Education Office, and Senior Consultant, Respiratory Medicine Service, Department of Paediatrics, KK Women's and Children's Hospital (KKH).

Beyond the hospital, Prof Chay's public healthcare posts include: Associate Designated Institutional Official, SingHealth Post-Graduate Medical Education; Professor, Duke-NUS Graduate Medical School and Yong Loo Lin School of Medicine, National University of Singapore; and Chair, Adolescent Health Steering Committee, Ministry of Health, Singapore.

Prof Chay has pledged her monetary award to the KKH Health Endowment Fund and SingHealth Duke-NUS Paediatrics Academic Clinical Program, in support of patients requiring financial assistance for medical treatment, and the advancement of clinical care, medical research and education.

BRINGING HOPE TO WOMEN AND CHILDREN IN NEED

KKH
HEALTH ENDOWMENT FUND

In celebration of 12 years of supporting women and children in need of financial assistance for medical treatment, KK Women's and Children's Hospital (KKH) unveiled the KKH Health Endowment Fund (KKHHEF) Donor Wall in July 2014.

"At KKH, we believe that every woman and child should have access to the best possible medical treatment and expertise," said Professor Kenneth Kwek, Chief Executive Officer, KKH.

"Since the KKHHEF began in 2002, we have had the unwavering support of many committed men, women and organisations whose generosity has made it possible to touch the lives of countless women, children and families in need."

Beneficiaries of the KKHHEF charity unveiling the donor wall at KKH.

"We gratefully acknowledge and honour them for their efforts in the fight against illness and disease through the KKHHEF Donor Wall."

GIVING PATIENTS A HEALTHIER, HAPPIER FUTURE

Each year, the KKHHEF provides financial assistance for medical treatment and intervention to over 500 women and children, many of whom suffer from chronic and critical conditions such as cancer, congenital abnormalities and issues associated with prematurity. The fund also supports education, research and disease prevention programmes that advance health for women and children.

To make a gift towards the KKHHEF, please contact Christine or Xian Hui at **+65 6394 2329 / 8439** or email **development@kkh.com.sg**.

Scan this code with your smartphone to find out more or visit **www.kkh.com.sg/GIVE**.

LIFE-SAVING HELP FOR YU ZHI

Yu Zhi was just three months old when he was diagnosed with chronic granulomatous disease – the same genetic immune system disorder that had caused the loss of his brother six years ago.

"We couldn't believe it," said Yu Zhi's mother.

Yu Zhi's treatment included injections, three times a week, costing $300. He also underwent a bone marrow transplant, which saw him hospitalised for two months.

Yu Zhi's parents, who had been experiencing financial constraints even before his diagnosis, had difficulty coping with his medical bills.

KKHHEF came to Yu Zhi's aid, providing financial assistance for medical supplies and special milk that the little boy needed to help meet his nutritional needs.

Today, while Yu Zhi still has to undergo drug therapy, he is a happy two-year-old.

"My greatest wish is for him to be able to run around and attend school like any normal child," says Yu Zhi's mother. "We are truly thankful to all the donors who give generously in aid of children like Yu Zhi."

If you would like to discontinue receiving Special Delivery or receive it at a different address, please contact us at **corporate.communications@kkh.com.sg** or **+65 6394 8048**.

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Contact Information:

KK Women's and Children's Hospital | 100 Bukit Timah Road, Singapore 229899 | Tel: +65 6-CALL KKH (6-2255 554) | Fax: +65 6293 7933 | Website: www.kkh.com.sg
Email: corporate.communications@kkh.com.sg | We are on facebook. 'Like' us for updates and news about KKH. | www.facebook.com/KKH.sg