Anesthetic considerations of a child with Down's syndrome

Essay

Submitted for partial fulfillment of
Master Degree in **Anesthesia**

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2014

بسم الله الرحمن الرحيم

وقل اعملوا فسيرى الله عملكم ورسوله والمؤمنون

صدق الله العظيم

سورة التوبة آية (١٠٥)

First, thanks are all due to **Allah** *for Blessing this work until it* has reached its end, as a part of his generous help throughout our life.

My profound thanks and deep appreciation to **Prof. Dr. Amir Ibrahim Salah**, *Professor of Anesthesia and Intensive Care, Faculty of Medicine, Ain Shams University for his great support and advice, his valuable remarks that gave me the confidence and encouragement to fulfill this work.*

I am deeply grateful to **Dr. Mohamed Nabil El Shafey,** *Assistant Professor of Anesthesia and Intensive Care,* Faculty of Medicine, Ain Shams university for adding a lot to this work by his experience and for his keen supervision.

I am also thankful to **Dr. Mohamed Sayed Shorbagy**, *Lecturer of Anesthesia and Intensive Care, Faculty of Medicine, Ain Shams University for his valuable supervision, co-operation and direction that extended throughout this work.*

I want also to thank ***my family*** *for supporting me throughout my life.*

Alia Mohamed Saad El Din

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List of abbreviations

AADI **:**anterior atlanto-dental interval

AAOD **:**anterior atlanto-occipital distance

AAP **:**American academy of pediatrics

Aptt **:**activated partial thromboplastin time

ARDS **:**adult respiratory distress syndrome

ASA **:**American society of anesthesiologists

ASD **:**atrial septal defect

AVSD **:**atrio-ventricular septal defect

CABG **:** coronary artery bypass graft

Co **:** carbon monoxide

Co_2 **:** carbon dioxide

CPAP **:**continuous positive airway pressure

CPB **:** cardio-pulmonary bypass

CRF **:**chronic renal failure

CSF **:** cerebrospinal fluid

CVP **:** central venous pressure

DS **:** down syndrome

ECG **:** electrocardiogaram

EF :ejection fraction

ESS **:**euthyroid sick syndrome

$ETCO_2$ **:**end tidal carbon dioxide

FEV1 **:**forced end expiratory pressure in first second

FRC **:**functional residual capacity

GERD **:**gastrointestinal reflux disease

GIT **:**gastrointestinal tract

IASD **:**inter auricular septal defect

ICU **:**intensive care unit

IVC **:**inferior vena cava

IVSD **:**inter ventricular septal defect

LV **:**left ventricle

NIBP **:**non invasive blood pressure

NICE **:** national institute for health and clinical excellence

NIPPV **:**non invasive positive pressure ventilation

NSAID **:** non steroidal anti-inflammatory drugs

OSA **:**obstructive sleep apnea

PACU **:**posa anesthetic care unit

PAH **:**pulmonary artery hypertension

PCWP **:**pulmonary capillary wedge pressure

PDA **:**patent ductus arteriosus

PEEP **:**positive end expiratory pressure

PONV **:**postoperative nausea and vomiting

PVD **:**pulmonary vascular disease

PVR **:**peripheral vascular resistance

REM **:**rapid eye movement

RV **:**right ventricle

SIRS **:**systemic inflammatory response syndrome

SVR **:**systemic vascular resistance

TEE **:**trans oesophageal ECHO

TOF **:**trachea-oesophageal fistula

TRH **:**thyrotropin releasing hormone

TSH **:**thyroid stimulating hormone

VC **:**vital capacity

VSD **:**ventricular septal defect

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Introduction

Down's syndrome, also known as Trisomy 21 is the commonest of congenital anomalies occurring 1 in 800 live births. It is characterized by dysmorphic facies. The incidence of Down's syndrome increases as the age of mother increases. The syndrome was first described by Dr. John Langdon Down in 1866. It is the best known chromosomal disorder in man, **(Yang et al., 2002)**.

The extra copy in chromosome 21 affects all the organ systems and results in a wide spectrum of phenotypic consequences resulting in the typical phenotype, mental retardation, hand anomalies and heart defects. Other features associated with Down's syndrome include macroglossia, microcephaly, endocardial cushion defects, ventricular septal defects, duodenal atresia, and atlanto-axial instability and supraglottic stenosis. All these result in unique sets of challenges to the anaesthesiologists (**James, 2006).**

Preoperative examination for patients with Down's syndrome should aim at a careful evaluation of organs and of systems which present malformations characteristic of this syndrome. The anesthetist has a very important role, since he must establish a relationship between patient status and perioperative risk (**Letterio et al., 2007).**

Anesthetic management is different in these patients due to the airway anomalies, congenital heart disease and endocrine anomalies. These patients are also prone to have hypothermia during surgery. Postoperative respiratory complications are also more common. All these considerations are to be taken care of during and after anesthesia and extra vigilance are to be maintained in these patients during the intraoperative and postoperative period **(Bhattarai et al., 2008).**

Aim of The Work

1- Discuss the anatomical and pathophysiological changes of Down's syndrome.

2- Preoperative assessment of child with Down's syndrome and the common procedures needed.

3- Focusing on the intraoperative techniques and modalities for safe anesthesia.

4- Reduction of postoperative complications.

Preoperative management of child with Down's syndrome

Down’s syndrome is the commonest chromosomal abnormality and is named after John Langdon Down, who described the syndrome in 1866.

Down’s syndrome is also referred to as Trisomy 21 due to the presence of an extra copy of chromosome number 21. It has an incidence of 1. 5 per 1000 live births **(Roizen, 2000; Carvalho, 2003).**

Chromosomal Changes Causing Down's syndrome:

Researches show that three types of chromosomal changes can lead to Down's syndrome:

Complete trisomy 21:

In this case, an error during the formation of the egg or the sperm results in either one having an extra chromosome, so after the egg and sperm unite, the resulting cells will also have three copies of chromosome 21. The complete extra copy of chromosome 21 is in all of the person's cells. Complete trisomy 21 is the cause of about 95% of Down's syndrome cases.

Mosaic trisomy 21:

Not every cell in the body is exactly the same. In about 1% of Down's syndrome cases, most of the cells in the body have the extra chromosome, but some of them don't. This is called "mosaicism. "

Mosaic trisomy 21 can occur when the error in cell division takes place early in development but after a normal egg and sperm unite. It can also occur early in development

when some cells lose an extra chromosome 21 that was present at conception.

The symptoms of someone with mosaic trisomy 21 may vary from those of someone with complete trisomy 21 or translocation trisomy 21, depending on how many cells have the extra chromosome.

Translocation trisomy 21:

In this type of chromosomal change, only part of an extra copy of chromosome 21 is in the cells. The extra part of the chromosome gets "stuck" to another chromosome and gets transmitted into other cells as the cells divide.

This type of change causes about 4% of Down 's syndrome cases. There are no distinct cognitive or medical differences between people with translocation trisomy 21 and those with complete trisomy 21 (**Parker et al., 2010).**

Multi-system problems in Down's syndrome:

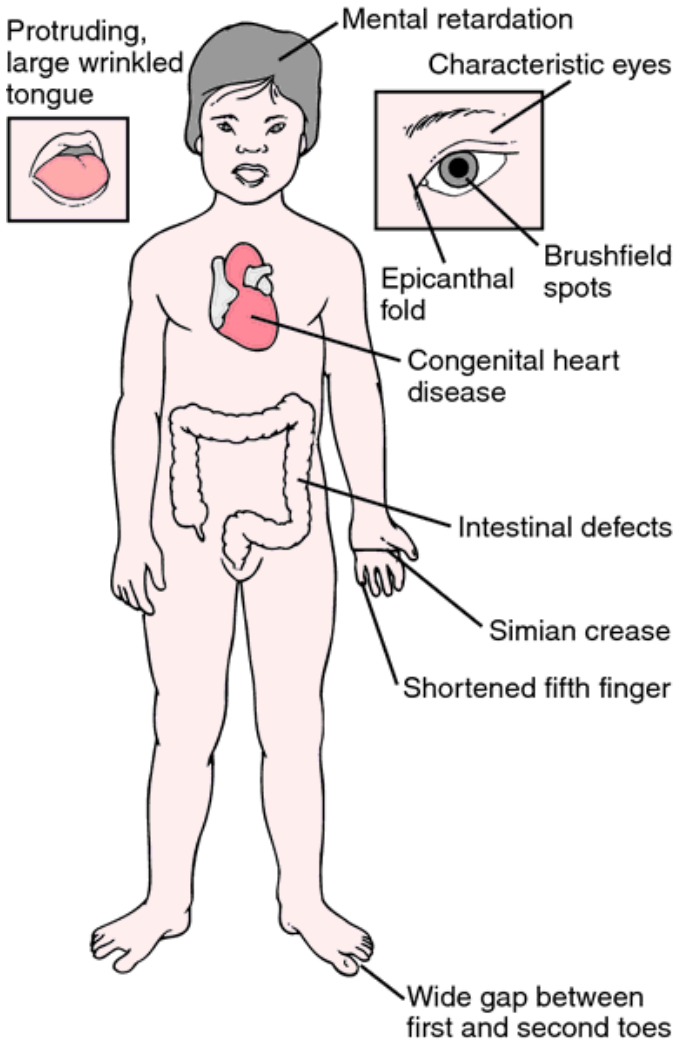

Fig. (1): Different features of Down's syndrome ***(Krishnan, 2009)***

General appearance :

- Small for age, generalized hypotonia, lax joints.
- Flat occiput, short neck, small low set ears, flattened nasal bridge with midface hypoplasia and protuding tongue.

- Prominent epicanthic folds (fold of skin of the upper eyelid covering the inner corner of the eye) and upward slanting eyes (Mongoloid slant).
- Brushfield spots (light coloured spots near the periphery of the iris).
- Single transverse palmar crease of the hand (Simian crease).
- Larger than normal space between the big and second toes (**Krishnan, 2009)**.

Cardiovascular system :

- Atrio-ventricular-septal defects (AVSD) (40%)
- Ventricular septal defects (VSD) (27%)
- Patent ductus arteriosus (PDA) (12%)
- Atrial septal defect (10%)
  • Tetralogy of Fallot (8%)
- Pulmonary vascular disease **(de Rubens et al., 2003)**.

Respiratory system :

- Recurrent respiratory tract infections, Sub-glottic stenosis, enlarged tongue, enlarged tonsils and adenoids and obstructive sleep apnea (**Letterio et al., 2007).**

Central nervous system :

- Developmental delay and moderate to severe mental retardation, microcephaly and epilepsy (5-10%) .

Skeletal system :

- Atlanto-axial instability: This is seen in about 15% of patients. The instability may be due to abnormality in the C1 vertebra (atlas) or C2 vertebra (axis), with laxity of the transverse ligament.