Ain Shams University
Faculty of Medicine
Department of Anesthesiology, Intensive Care and Pain Management

Anesthetic management of pediatric patient with congenital hyperinsulinism undergoing pancreatectomy

An Essay Submitted for Partial Fulfillment of Master Degree in Anesthesia

By

Haitham Abd el Reheem Nawar
M.B.B.Ch. (Ain Shams University)
Under Supervision of

PROF.DR.HANY MOHAMED EL ZAHABY
Professor of Anesthesia, Intensive Care and Pain Management
Faculty of Medicine – Ain Shams University

DR.ABD EL AZIZ ABD ALLAH ABD EL AZIZ
Lecturer of Anesthesia, Intensive Care and Pain Management
Faculty of Medicine – Ain Shams University

DR.WAHBA ZAKARIA WAHBA
Lecturer of Anesthesia, Intensive Care and Pain Management
Faculty of Medicine – Ain Shams University

Faculty of Medicine
AinShamsUniversity
2015

بسم الله الرحمن الرحيم

وَقُلِ اعْمَلُوا فَسَيَرَى اللَّهُ عَمَلَكُمْ وَرَسُولُهُ وَالْمُؤْمِنُونَ

صدق الله العظيم

سورة التوبة آية (105)

ACKNOWLEDGEMENT

*First, I would like to express my deepest gratitude and thanks to **Allah** whose magnificent help was the main factor in accomplishing this work.*

*No words can adequately express my sincere gratitude and great appreciation to **Prof. Dr. Hany Mohamed El Zahaby,** Professor of Anesthesia and I.C.U., faculty of medicine; Ain shams University, for his encouragement, helpful supervision and guidance.*

*I am also expressing my deepest thanks to **Dr.Abd el aziz Abdallah Abd el aziz,** Lecturer of Anesthesia and I.C.U., faculty of medicine; Ain shams University, for his continuous advice, valuable assistance and efforts in this work.*

*I would also like to express my sincerest thanks and deep appreciation to **Dr. Wahba Zakaria Wahba,** Lecturer of Anesthesia and I.C.U., faculty of medicine; Ain shams University, for his valuable supervision and kind instructions.*

*I am extremely sincere to **my family** who stood beside me throughout this work giving me their support.*

Haitham Abd el reheem Nawar

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List of Abbreviations

B-cells	Beta cells of the pancreas
BP	Blood pressure
BWS	Beckwith Wiedemann syndrome
CHI	Congenital hyperinsulinism
C-peptide	Connecting peptide
ECG	Electro-cardiogram
ETT	Endotracheal tube
GHD	Glutamate dehydrogenase
HbA1C	Glycated hemoglobin
HI/HA	Hyperinsulinism Hyperammonemia
IRS	Insulin resistance syndrome
K-ATP	Inward rectifying potassium cells
LGA	Low for gestational age
MEN	Multiple endocrine neoplasia
OR	Operating room
PACU	Post anesthesia care unit
PET	Positron emission tomography
PHHI	Persistent hyperinsulinemic hypoglycemia
PP cells	Pancreatic polypeptide cells of pancreas
SGA	Small for gestational age
Spo2	Peripheral capillary oxygen saturation
SUR1	Sulfonyl urea receptor
UCP	Uncoupling protein
URI	Upper respiratory infection

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Introduction and Aim of the Essay

Introduction

Congenital hyperinsulinism (CHI) is the most common cause of persistent hypoglycemia in infants and children. In most affected infants, CHI is caused by a specific genetic defect that results in the altered expression of pancreatic beta cells causing unregulated oversecretion of insulin. Infants with CHI may have either focal or diffuse abnormalities of the pancreatic b-cells. Both forms of CHI manifest as hypoglycemia, usually in the early newborn period. Focal disease can be treated effectively with surgical resection of the affected area, resulting in a total cure or rendering the patient amenable to medical management. Most children with diffuse disease are unresponsive to medical therapy, and require near-total pancreatectomy ***(Olga et al, 2012).***

An estimated incidence of 1 in 50,000 live births in a random-mating United States population has been reported. Worldwide, the incidence may be as high as 1 in 2500 live births in populations with high rates of consanguineous unions. Patients with CHI usually present between birth and age 18 months, with most cases diagnosed shortly after birth ***(Kapoor et al, 2013).***

The main aim of anesthetic management is to prevent hypoglycemia until resection of the pancreas and the control of rebound hyperglycemia soon after resection. Various approaches have been described, including use of "artificial pancreas", which continuously monitors plasma glucose and delivers glucose or insulin to maintain a predetermined glucose level. But, it is not much in use due to its cost and complexity ***(Manabe et al, 2009).***

Pancreatectomy is an urgent surgery that is performed to protect the brain. It is a safe procedure that is well tolerated in infants and children, and successful anesthetic management depends on fluid management, to maintain normoglycemia in spite of changing glucose levels, and to maintain hemodynamic stability during the perioperative period. This procedure is always a challenge to many anesthesiologists ***(Manabe et al, 2001).***

Another suggested approach is continuous infusion of 10% glucose and every 15-min monitoring of blood glucose levels to maintain plasma glucose in the range of 100–150 mg/dL. This method helps to detect sudden hypo or hyperglycemia ***(Yamashita et al, 2006).***Other groups

recommended this frequency of sampling every 30 min ***(Akhtaruzzaman et al, 2008).***

Aim of the Essay

The aim of this essay is to highlight the epidemiology and pathophysiology of congenital hyperinsulinism and the preoperative preparation, intraoperative anesthetic management and postoperative care of pediatric patient with congenital hyperinsulinism undergoing pancreatectomy.

Epidemiology and Pathophysiology of Congenital Hyperinsulinism

Epidemiology

Persistent hyperinsulinemic hypoglycemia of infancy (PHHI) represents the most common cause of hyperinsulinism in neonates. Currently, many authors prefer the term congenital hyperinsulinism (CHI)
(Prabudh and Subhasis, 2012).

An estimated incidence of 1 in 50,000 live births in a random-mating United States population has been reported. Worldwide, the incidence may be as high as 1 in 2500 live births in populations with high rates of consanguineous unions ***(Kapoor et al, 2013).***

Patients with CHI usually present between birth and age 18 months, with most cases diagnosed shortly after birth. Cases of adult-onset forms of CHI are rare but well documented. The overall male-to-female ratio is 1.3:1.The diffuse form of CHI has a male-to-female ratio of 1.2:1.Focal lesions form of CHI has a male-to-female ratio of 1.8:1 ***(Robert et al, 2010).***

Congenital hyperinsulinism can have different inheritance patterns, usually depending on the form of the

condition. At least two forms of the condition have been identified. The most common form is the diffuse form, which occurs when all of the beta cells in the pancreas secrete too much insulin. The focal form of congenital hyperinsulinism occurs when only some of the beta cells over-secrete insulin ***(James et al, 2009).***

Most often, the diffuse form of congenital hyperinsulinism is inherited in an autosomal recessive pattern, which means both copies of the gene in each cell have mutations. The parents of an individual with an autosomal recessive condition each carry one copy of the mutated gene, but they typically do not show signs and symptoms of the condition ***(James et al, 2009).***

Less frequently, the diffuse form is inherited in an autosomal dominant pattern, which means one copy of the altered gene in each cell is sufficient to cause the disorder ***(James et al, 2009).***

The inheritance of the focal form of congenital hyperinsulinism is more complex. For most genes, both copies are turned on (active) in all cells, but for a small subset of genes, one of the two copies is turned off (inactive).