A Practical Handbook on Pediatric Cardiac Intensive Care Therapy

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Foreword

The most sophisticated way of teaching medicine is to transfer knowledge, experience, and skills directly into newly built medical centers equipped with modern devices. A new generation of doctors is coming to the fore in neonatal cardiovascular intensive care, and it is necessary for them to start by successfully applying theoretical knowledge. This book provides patient-oriented approaches to the interdisciplinary specialists involved in decision-making at the point of care and to those who need to be able to understand and manage the concrete situations in neonates and small children with congenital heart defects.

The core value of this book for beginners is its handover of clinically relevant information while promoting the fundamentalist's style of thinking about physiology and organ function in different congenital heart defects. Such an approach drives an understanding of the forthcoming problems and how to manage them. Besides understandable and detailed descriptions of specific pitfalls and special situations relating to the diagnosis, intervention, and medical treatment of each distinctive defect, this practical handbook focuses on general daily practice and bedside medicine for doctors and nurses: easy to understand, straightforward to implement, and result-oriented. Its chapters fill the informational gaps in cardiopulmonary interactions to manage the multiple hemodynamic situations that can arise.

Doctors in Russia, Eastern European, and Asia are confronted with difficult-to-manage neonates and complex technologies in the absence of advanced knowledge in both the operating theater and the ICU. For these reasons, newly built centers suffer from high mortality and complication rates even in simple cases and are therefore only able to treat a limited spectrum of patients in urgent need of cardiac surgery. This book enables its readers to recognize in advance the signs of approaching emergency situations and adapt to the situation in a timely manner or obtain the appropriate help. For each patient on the ICU, it provides a broad knowledge base to understand what is happening, foresee complications, and react quickly to arising problems.

During their medical missions to the Neonatal Center of St. Petersburg State Pediatric Medical University where a new pediatric heart program is being developed, Dietrich Klauwer and Christian Jux provided one-on-one teaching to the local specialists. Thanks to the first German version of this book, it was possible to introduce training systems for beginners on pediatric cardiac ICU in St. Petersburg, Russia. This book details ways to manage complex problems in a pragmatic,

concrete, and experience-based way. The authors achieved the goal of creating a work with proven effectiveness that has become the cornerstone in the practical education of next interdisciplinary generation of cardiovascular critical care specialists.

January 2018

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Foreword

Over the past decades, the care for children with congenital heart disease has seen significant change. The number of operations and the complexity of surgery have increased. This included a move from initial palliation to early repair of several structural lesions. More challenging surgical approaches warranted more specialized pediatric cardiac critical care units which were established at larger centers in Europe and North America, leading to better outcomes but also to a need for more trained and knowledgeable nurses and physicians in this area of patient care. A new pediatric subspecialty was emerging – but sometimes without the usual established training programs. Gaining appropriate knowledge may become a challenge. For the beginner in the field of pediatric cardiac critical care, everything is new. Being exposed to (different) terminologies of congenital heart disease, making the correct diagnosis and treatment plan is sometimes challenging. In the interests of the children who need advanced care on the critical care unit, the entire team – physicians and nurses – need to understand both normal and relevant physiology of the underlying defect along with preoperative and post-intervention hemodynamics.

The great advantage of this book is that it serves the learners, while taking command as reference for advanced practitioners in that it bridges many areas, and also provides detailed problem-solving.

Without repeating the table of contents, a couple of important chapters should be highlighted, emphasizing well-explained ventilation strategies for different situations and frequently used medications (antibiotics, inotropes, vasopressors, etc.). Analgesia and sedation pathways for early extubation or prolonged ventilation are described, and a pro/con chapter for fast track is included. In addition, the authors cover important other areas like structured handover, nutrition, pulmonary hypertension, and mechanical support. Well-designed illustrations and tables provide an instant overview.

With a consistent approach that was long practiced and nurtured, the authors of this book achieved the goal of aligning all the aforementioned topics into a German version which was published several years ago and highly appreciated. It was no wonder that the first edition was rapidly sold.

I congratulate the team and the Springer-Verlag on this tremendously important project. Given that a second German edition has already been published, minor problems have been ironed out and newer treatment algorithms are included. The English edition is targeted to an international audience, including countries that are

in the process of developing cardiac surgical centers and, therewithin, nursing, pediatric intensivists, cardiologists, anesthesiologists, and surgeons who are bothin training and early practice. Rather than rigid instruction, this book truly serves as a guideline or curriculum whereupon programs can build – and I believe it will enable the next generation of caregivers in our field to reach the next level.

January 2018

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Preface

The motivation to have the second German edition of our well-received practical handbook on pediatric cardiac intensive care translated into English was to impart to the global medical community our mostly pathophysiologically and experience-oriented methods of managing children with cardiac pathologies. This intention emerged from three directions.

Need

During many travels to foreign countries in Eastern Europe, Asia and Africa on missions to help departments initiate or develop state-of-the-art pediatric heart surgery programs, it became evident that the greatest deficits in the postoperative therapy of children lie in the lack of clear, comprehensible and actionable strategies. Established protocols and workflows for clinical assessment, preoperative diagnostics through therapeutic management to planning and performing surgeries in the hands of a well-coordinated team are not in place in many developing centres.

Here, the most obvious need was how to reproduce for and share with others the combined experience gained over decades. This need for knowledge transfer applied to the clinical assessment of patients, their organ functions in critical situations, how to anticipate disease courses, teach others how to deploy modern equipment and administer pharmacological treatments in the best available way.

With the present handbook, the authors hope to create a pathophysiological understanding for the processes, problems and complications routinely encountered on a pediatric cardiac ICU so that options for action can be found and ways to find solutions made transparent.

Insofar as we wanted to do justice to the academic dispute about “evidence”, we felt a handbook structure seemed more purposeful for explaining how to achieve successful therapeutic outcomes on an individualized basis. That is only one of the reasons why this handbook does not claim to present the reader with elaborate literature searches on theoretically best practices. Rather, it combines the many authors’ multiple years of experience in establishing self-verified, innovative and advanced clinical methods that fit our own effective concepts, while also taking the currently relevant literature into account in the true sense of evidence-based medicine.

Ambition

The idea to reshape the mould of an introductory guidance intended for the Giessen pediatric ICU into a handbook of critical care on a pediatric ICU was born in the year 2011. After publication of the first German edition and its unexpectedly high acceptance, the authors decided to write a second, revised and extended version. For this second issue, the authors were able to inspire other renowned authoritative contributors and with these co-authors broaden the book's scope. To this new version, we added important related topics that are pivotal to enriching the readers' knowledge about the hemodynamics of different heart defects and the postoperative circulatory changes occurring with and without the use of respirators.

Our desire was to satisfy the unmet need described above and to compete with other English works on the subject. Convinced about its concept and committed to an implementation that goes against the grain of the conventionally practiced way of sharing medical knowledge, we aim to open up the contents of this handbook to a worldwide public working in the field of pediatric cardiology.

Opportunity

The real opportunity to be able to turn this ambitious goal of a first English edition into reality was enabled by Getinge thanks to their sponsorship of a professional medical translation. The authors also feel highly fortunate to have this work published by the distinguished Springer Verlag with their affiliated sales channels of a global distribution network.

Above and beyond that appreciation, my thanks go to all of those who participated with heart, hand and mind in realization of this project: to the Eurasia Heart Foundation for their exemplarily contribution to the original idea, to Ms. Deborah A. Landry for her infinite patience during the translation process, to Ms. Katja Kassem for the figures, to the German publisher Deutscher Ärzteverlag GmbH for transferring the rights as well as to all the authors and their families for their support in getting this book out alongside their routine clinical work.

For the authors Dietrich Klauwer, Feb. 2018

Singen, Germany — Dietrich Klauwer
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My conflict of interest is as follows: R. Zimmermann is an employee of Actelion Pharmaceuticals, Switzerland.

Contents

Part II Management of Specific Pediatric Cardiac Problems

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List of Abbreviations

AA	Amino acid(s)
AB	Antibiotics
Ab	Antibodies
ABB	Acid-base balance
ABP	Arterial blood pressure
ACC	Acetylcysteine
ACE	Angiotensin-converting enzyme
Acetyl-CoA	Acetyl coenzyme A
ACT	Activated clotting time
ACTH	Adrenocorticotropic hormone
AdC	Adenylate cyclase
ADH	Antidiuretic hormone
ADP	Adenosine diphosphate
ADR	Adverse drug reaction
aEEG	Amplitude-integrated electroencephalography
AG	Anion gap
AKI	Acute kidney injury
ALI	Acute lung injury
ALS	Advanced Life Support
ALT	Alanine aminotransferase
ANP	Atrial natriuretic peptide
AP	Aortopulmonary
AP shunt	Aortopulmonary shunt
APC	Activated protein C
APh	Alkaline phosphatase
ARDS	Acute respiratory distress syndrome
AS	Aortic stenosis
ASD	Atrial septal defect
ASO	Arterial switch operation
ASS	Acetylsalicylic acid
AST	Aspartate aminotransferase
AT III	Antithrombin III
ATG	Antithymocyte globulin
ATP	Adenosine triphosphate

AVB, AV block	Atrioventricular block
$avDO_2$	Arteriovenous oxygen difference
AVNRT	Atrioventricular nodal reentry tachycardia
AVRT	Atrioventricular reentrant tachycardia
AVSD	Atrioventricular septal defect
AWMF	Association of the Scientific Medical Societies in Germany
AZA	Azathioprine
bid	Twice a day (“bis in die”)
BAL	Bronchoalveolar lavage
BAP	Balloon angioplasty
BB	Buffer base
BC	Blood culture
BE	Base excess
BEecf	Base excess of extracellular fluid
BG	Blood gas
BG	Blood group
BGA	Blood gas analysis
Bili	Bilirubin
BIPAP	Biphasic positive airway pressure
BIS	Bispectral index
BLD	Blood leak detector
BLS	Basic life support
BNP	Brain natriuretic peptide
BP	Blood pressure
BPD	Bronchopulmonary dysplasia
BS	Blood sugar
BSA	Body surface area
BT Shunt	Blalock-Taussig Shunt
BW	Body weight
Ca	Calcium
cAMP	Cyclic 3’,5’-adenosine monophosphate
CaO_2	O_2 content of arterial blood
CAPD	Continuous ambulatory peritoneal dialysis
CBF	Cerebral blood flow
CPB	Cardiopulmonary bypass
CC	Creatinine clearance
CCB	Calcium channel blocker
CCT	Cranial computed tomography
CCT	Aortic cross-clamp time
CDH	Congenital diaphragmatic hernia
Cdyn	Dynamic compliance
CF	Cystic fibrosis
cGMP	Cyclic guanosine monophosphate
CH	Charrière
CI	Cardiac Index

CID	Continuous intravenous drip infusion
CK	Creatine kinase
CK-MB	Creatine kinase-muscle/brain
CM	Contrast medium
CMV	Cytomegalovirus
CN	Cyanide
CNS	Central nervous system
CO (Q)	Cardiac output
CO_2	Carbon dioxide
CoA	Coarctation of the aorta
COX	Cyclooxygenase
CPAP	Continuous positive airway pressure
CPB	Cardiopulmonary bypass (heart-lung machine)
CPP	Cerebral perfusion pressure
CPR	Cardiopulmonary resuscitation
CPVT	Catecholaminergic polymorphic ventricular tachycardia
CRC	Concentrated red cells
CrCl	Creatinine clearance
CrP	C-reactive protein
CRRT	Continuous renal replacement therapy
CsA	Cyclosporine A
CSD	Coronary sinus defect
CT	Clotting time
CTEPH	Chronic thromboembolic pulmonary hypertension
CVC	Central venous catheter
$CvCO_2$	Venous concentration of carbon dioxide
CVP	Central venous pressure
CVVHDF	Continuous venovenous hemodiafiltration
D	Dislocation
Da	Dalton
DA	Duration of action
DAP	Diastolic arterial pressure
DCM	Dilative cardiomyopathy
DHCA	Deep hypothermic circulatory arrest
DIC	Disseminated intravascular coagulation
DILV	Double inlet left ventricle
DKS	Damus-Kaye-Stansel procedure
dl	Deciliter (100 ml)
DNA (S)	Deoxyribonucleic acid
DO_2	Oxygen delivery
DORV	Double outlet right ventricle
dP, ΔP	Pressure change (delta P)
DPG	Diphosphoglycerate
DSO	German Organ Transplantation Foundation
d-TGA	Dextro-transposition of the great arteries

dV	Volume change (delta V)
dyn	dyne, unit of force equal to 10^{-5} newton
E-lyte	Electrolyte
EBV	Epstein-Barr virus
ECC	Extracorporeal circulation
ECLS	Extracorporeal life support
ECMO	Extracorporeal membrane oxygenation
ECS	Extracellular space
EDTA	Ethylenediaminetetraacetic acid
EDV	End-diastolic volume
EF	Ejection fraction
ELSO	Extracorporeal Life Support Organization
EMA	European Medicines Agency
ERA	Endothelin receptor antagonist
ERC	European Resuscitation Council
ERO_2	O_2 extraction ratio
ESBL	Extended-spectrum beta-lactamase
ESC/ERS	European Society of Cardiology/European Respiratory Society
ET	Eurotransplant
ET-1	Endothelin-1
ET-A, ET-B	Endothelin-A, Endothelin-B
$etCO_2$	End-tidal CO_2
F	French (scale for denoting the size of catheters)
FAT	Focal atrial tachycardia
FDA	United States Food and Drug Administration
FDP	Fibrin degradation products
$FECO_2$	Fraction of end tidal CO_2
FeNa	Fractional excretion of sodium
FFA	Free fatty acids
FFP	Fresh frozen plasma
FIB	Fibrinogen
FiO_2	Fraction of inspired oxygen
FPE	First-pass effect
FRC	Functional residual capacity
FS	Fraction shortening (shortening fraction)
FS	Fractional shortening
FV	Factor V
FVIII	Factor VIII
G	Gauge
GABA	Gamma-aminobutyric acid
GABAergic	Gamma-aminobutyric acid-ergic
GCS	Glasgow Coma Scale
G-CSF	Granulocyte colony-stimulating factor
GFR	Glomerular filtration rate
GH	Growth hormone

GHB	Gamma-hydroxybutyric acid
GI	Gastrointestinal
h	Hour(s)
HA	Human albumin
Hb	Hemoglobin
HbO_2	Oxyhemoglobin
HBsAG	Hepatitis B surface antigen
HBV	Hepatitis B virus
HCO_3^-	Hydrogen carbonate
Hct	Hematocrit
HCV	Hepatitis C virus
HD	Hemodialysis
HDF	Hemodiafiltration
HES	Hydroxyethyl starch
HF	Hemofiltration
HFOV	High-frequency oscillation ventilation
HIF	Hypoxia-induced factor
HIT	Heparin-induced thrombocytopenia
HIT-II	Heparin-induced thrombocytopenia type II
Hct	Hematocrit
HLA	Human leukocyte antigen
HLH	Hypoplastic left heart
HLHS	Hypoplastic left heart syndrome
HLM	Heart-lung machine
HOCM	Hypertrophic obstructive cardiomyopathy
HPT	Hyperparathyroidism
HPV	Hypoxic pulmonary vasoconstriction
HR	Heart rate
HTx	Heart transplantation
i.m.	Intramuscular
i.o.	Intraosseous
i.v.	Intravenous
IAA	Insulin autoantibodies
IABP	Intra-aortic balloon pump
IAP	Intraabdominal pressure
IART	Intra-atrial reentrant tachycardia
ICB	Intracranial bleeding
ICD	Implantable cardioverter-defibrillator
ICP	Intracranial pressure
ICU	Intensive care unit
ID	Internal diameter
IGF	Insulin-like growth factor
IgG	Immunoglobulin G
INR	International normalized ratio
IP receptor	Prostacyclin receptor (I-Prostanoid)

iPAH	Idiopathic pulmonary arterial hypertension
ISA	Intrinsic sympathomimetic activity
ISHLT	International Society for Heart and Lung Transplantation
ISTA	Aortic isthmus stenosis
I-time	Inspiratory time
IU	International unit
IVC	Inferior vena cava
IVH	Intraventricular hemorrhage
IVIG	Intravenous immunoglobulin
IVS	Intact ventricular septum
J	Joule
JET	Junctional ectopic tachycardia
K	Potassium
KCl	Potassium chloride
Kg	Kilogram
KUSS	Childhood Discomfort and Pain Scale (German: Kindliche Unbehagen- und Schmerz-Skala)
L/R shunt	Left/right shunt
LA	Left atrium
LAD	Left anterior descending
LAP	Left atrial pressure
LCO	Low cardiac output
LDH	Lactate dehydrogenase
LI	Liver insufficiency
LIP	Lower inflection point
LMA	Laryngeal mask airway
LMWH	Low molecular weight heparin
Lp	Lipoprotein
LP	Lumbar puncture
LPA	Left pulmonary artery
LPOHV	Left-persisting upper vena cava
LPR	Lactate-pyruvate ratio
LPS	Lipopolysaccharide
LT	Long-term
l-TGA	Levo-transposition of great arteries
LTx	Liver transplantation
LuFu	Lung function
LV	Left ventricle
LVEDD	Left ventricular end-diastolic diameter
LVEDP	Left ventricular end-diastolic pressure
LVOT	Left ventricular outflow tract
LVOTO	Left ventricular outflow tract obstruction
MA	Maximum amplitude
MA	Maximum amplitude in thromelastography

MABP	Mean arterial blood pressure
MAC	Minimum alveolar concentration
MAPCA	Major aortopulmonary collateral artery
MAPSE	Mitral annular plane systolic excursion
mbar	Millibar
MC	Microcirculation
MCF	Maximum clot firmness
mcg	Microgram
mEq	Milliequivalent
Met-Hb	Methemoglobin
Mg	Magnesium
mg%	Milligrams percent
MI	Mitral insufficiency
MIBI	Microbiology
MIC	Minimum inhibitory concentration
min	Minute(s)
ML	Maximum lysis
µm	Micrometer
MMF	Mycophenolate mofetil
mmHg	Millimeters of mercury
mmol	Millimol
MNF	Multiresistant nonfermenters
mo	Month(s)
MOF	Multi-organ failure
Mol	Mol
mosmol	Milliosmol
MRGN	Multi-resistant gram-negative
MRSA	Methicillin-resistant *Staphylococcus aureus*
MRSE	Methicillin-resistant *Staphylococcus epidermidis*
MRT	Magnetic resonance tomography
MST	Mitral stenosis
MTHFR	Methylenetetrahydrofolate reductase
MTX	Methotrexate
mU	Milliunits
MUF	Modified ultrafiltration
MV	Minute volume
mV	Millivolt
Na	Sodium
NaBi	Sodium bicarbonate
NAC	N-acetylcysteine
NAD	Nicotinamide adenine dinucleotide
NADH	Nicotinamide adenine dinucleotide (reduced)
NAPQI	N-acetyl-P-benzoquinone imine
NBP	Non-bicarbonate buffer

NEC	Necrotizing enterocolitis
NH_3	Ammonia
NIRS	Near-infrared spectroscopy
NMDA	N-methyl-D-aspartate
NO	Nitrogen oxide
NOAD	New oral anticoagulant drugs
NSAID	Nonsteroidal anti-inflammatory drug
NSE	Neuron-specific enolase
NYHA	New York Heart Association
O	Obstruction
O_2	Oxygen
OLT	Open Lung Tool
OP	Operation
ORT	Orthodromic reentry tachycardia
P	Phosphorus
p.o.	Per os
PA	Pulmonary artery
PAC	Pulmonary artery catheter
PaCO2	Arterial partial pressure of carbon dioxide
PAH	Pulmonary arterial hypertension
PAH-CHD	Pulmonary arterial hypertension associated with congenital heart disease
PAI	Plasminogen activator inhibitor
PALS	Pediatric advanced life support
PAM	Postaggression metabolism
PaO_2	Reduced oxygen tension
PAP	Pulmonary artery pressure
PAPm	Mean pulmonary artery pressure
PAPVR	Partial anomalous pulmonary venous return
PAS	Postaggression syndrome
PAT	Pulmonary atresia
PAWP	Pulmonary arterial wedge pressure
PBF	Pulmonary blood flow
PBP	Pre-blood pump
PC	Platelet concentrate
PC	Pressure control
PCA	Patient-controlled analgesia
PCH	Pulmonary capillary hypertension
PCM	Paracetamol
PCO_2	Partial pressure of carbon dioxide
PCR	Polymerase chain reaction
PCT	Procalcitonin
PCWP	Pulmonary capillary wedge pressure
PD	Peritoneal dialysis
PDA	Patent ductus arteriosus

PDE	Phosphodiesterase
PDE5i	Phosphodiesterase-5 inhibitor
PdGF	Platelet-derived growth factor
PDR	Pulmonary vascular resistance
PE	Pulmonary embolisms
PEA	Pulseless electrical activity
PEEP	Positive end-expiratory pressure
PEG	Percutaneous endoscopic gastrostomy
PET	Positron emission tomography
$PetCO_2$	Partial end-tidal carbon dioxide tension
PF	Platelet factor
PF4	Platelet factor 4
PFC	Persistent fetal circulation
PFK	Phosphofructokinase
PFO	Patent foramen ovale
PG	Prostaglandin
PGH_2S	Prostaglandin H_2 synthase
PGI_2	Prostaglandin I_2 (prostacyclin)
pH	hydrogen ion (H+) concentration (acidity) of a solution, ranging from 0 to 14
PH_2O	Hydrostatic pressure
PHT	Pulmonary hypertension
PIP	Peak pressure or positive inspiratory pressure
PJRT	Persistent junctional reciprocating tachycardia
pKa	Acid dissociation constant
PLS	Pediatric Life Support
PM	Pacemaker
PMN	Polymorphonuclear neutrophils
PN	Premature neonate
PaO_2	Partial pressure of oxygen
POCT	Point-of-care testing
POD	Postoperative day
PPHN	Persistent pulmonary hypertension of the newborn
ppm	Parts per million
PPN	Partial parenteral nutrition
PPSB	Prothrombin, proconvertin, Stuart-Prower factor, antihemophilic factor B
PPV	Positive pressure ventilation
PRA	Panel reactive antigen
PRIS	Propofol infusion syndrome
PRVC	Pressure-regulated volume control
PS	Pressure support
PST	Pulmonary stenosis
PSV	Pressure support ventilation
PSVT	Paroxysmal supraventricular tachycardia
PTA	Persistent truncus arteriosus

PTFE	Polytetrafluorethylene
PTH	Parathyroid hormone
PtO_2	Tissue oxygen partial pressure
PTT	Partial thromboplastin time
PV	Pulmonary vein
PvCO2	Venous partial pressure of carbon dioxide
PVL	Periventricular leukomalacia
PVO	Pulmonary venous obstruction
PvO_2	Low oxygen partial pressure
PVR	Pulmonary vascular resistance
PVRI	Pulmonary vascular resistance index
Qp	Ratio of pulmonary
Qp/Qs	Ratio of pulmonary-to-systemic blood flow
Qs	Systemic perfusion
RA	Right atrium
RAAS	Renin-angiotensin-aldosterone system
RACE	Repetitive alveolar collapse and expansion
RAP	Right atrial pressure
RBF	Renal blood flow
RDS	Respiratory distress syndrome
Rea	Reanimation
RI	Renal impairment
RMV	Respiratory minute volume
RNA	Ribonucleic acid
ROSC	Return of spontaneous circulation
ROTEM	Rotational thromboelastometry
Rp	Pulmonary vascular resistance
RPA/LPA	Right/left pulmonary artery
rpm	Revolutions per minute
RPP	Renal perfusion pressure
RQ	Respiratory quotient
RR	Respiratory rate
RRT	Renal replacement therapy
$rSCO_2$	Regional cerebral oxygen saturation
r-tPA	Recombinant tissue plasminogen activator
RV	Right ventricle
RVEDP	Right ventricular end-diastolic pressure
RVOT	Right ventricular outflow tract
RVOTO	Right ventricular outflow tract obstruction
RVP	Right ventricular pressure
S	Sieving coefficient
SA block	Sinoatrial block
SaO_2	Arterial oxygen saturation
SAP	Systolic arterial pressure, supra-arterial pressure

SBE	Standard base excess
SCD	Sudden cardiac death
$ScvO_2$	Central venous saturation
sec	Second(s)
SF	Surfactant
sGC	Soluble guanylate cyclase
SIADH	Syndrome of inappropriate antidiuretic hormone secretion
SID	Strong ion difference
SIDS	Sudden infant death syndrome
SIMV	Synchronized intermittent mandatory ventilation
SIPPV	Synchronized intermittent positive pressure ventilation
SIRS	Systemic inflammatory response syndrome
SO_2	Oxygen saturation in general
SpO_2	Saturation of peripheral oxygen
$SpvO_2$	Pulmonary venous saturation
SR	Sinus rhythm
STB	Standard bicarbonate
SV	Stroke volume
SVC	Superior vena cava
SVD	Sinus venosus defect
SVES	Supraventricular extra systole(s)
SvO_2	Venous saturation
SVR	Systemic vascular resistance
SVT	Supraventricular tachycardia
T3	Triiodothyronine
T4	Thyroxin
TAC	Tacrolimus
TAC	Truncus arteriosus communis
TAPSE	Tricuspid annular plane systolic excursion
TAPVR	Total anomalous pulmonary venous return
TAT	Tricuspid atresia
Tc	Transcutaneous (saturation)
TC	Time constant
TCO_2	Total carbon dioxide
TCPC	Total cavopulmonary connection
TEE	Transesophageal echocardiography
TEG	Thrombelastography
TEI	Myocardial performance index
Temp	Temperature
TFA	Total fluid amount
TFPI	Tissue factor pathway inhibitor
TGA	Transposition of great arteries
TGF	Transforming growth factor
TI	Tricuspid insufficiency

Ti/Te	Inspiratory time/expiratory time
TIVA	Total intravenous anesthesia
TMP	Transmembrane pressure
TNI	Troponin I
TOF	Tetralogy of Fallot
TOR	Target of Rapamycin
tPA	Tissue plasminogen activator
TPG	Transpulmonary pressure gradient
TPN	Total parenteral nutrition
TPG	Transpulmonary gradient
TPR	Tubular phosphate reabsorption
TRALI	Transfusion-associated acute lung injury
TRIS	Tris(hydroxymethyl)aminomethane buffer ($C_4H_{11}NO_3$)
TT	Thrombin time
TU	Tumor
TÜV	German Technical Inspection Association
TV	Tricuspid valve
U	Units
UDP-GT	Uridine diphosphate glucuronosyltransferase
UFH	Unfractionated heparin
UIP	Upper inflection point
UTI	Urinary tract infection
UTS	Ullrich-Turner syndrome
UVC	Umbilical venous catheter
V/Q	Ventilation-perfusion ratio
VA	Alveolar ventilation
VAD	Ventricular assist device
$vaDCO_2$	Venoarterial carbon dioxide difference
VAP	Ventilator-associated pneumonia
VCO2	Carbon dioxide output
VD	Volume dead space
VEGF	Vascular endothelial growth factor
VES	Ventricular extra systole(s)
VF	Ventricular fibrillation
VILI	Ventilator-induced lung injury
Vmax	Maximum velocity
VO_2	Oxygen consumption
VP shunt	Ventriculoperitoneal shunt
VRE	Vancomycin-resistant enterococci
VSD	Ventricular septal defect
Vt	Tidal volume
VT	Ventricular tachycardia
VV	Venovenous
vWF	von Willebrand factor
WBS	Williams-Beuren syndrome

WL	Week of life
WPW	Wolff-Parkinson-White syndrome
WU	Wood units
YC	Young children
yr	Year(s)

Important General Preliminary Remarks

Dietrich Klauwer

Organization

This book is addressed to the beginner in pediatric intensive care and pediatric cardiac intensive care and in its general section is intended to communicate the principles of practical management of the patient in a pediatric intensive care unit. In the authors' view, as well as a basic knowledge of the functioning and monitoring of the different organ systems, this also includes knowledge of the individual patient's problems.

This knowledge should provide the newcomer with a clear framework within which he or she can rapidly gain confidence in his or her management of frequently extremely severely ill patients, despite the complexity of the setting.

In order to be able to provide rapid help in an emergency and to obtain assistance, it is vital to know the logistics of the site and to have key data on all patients at hand. In addition to an *understanding of the monitoring unit,* this also includes knowledge about the handling of suction systems and the ventilation bags adapted to different patient sizes and the operation of ventilation devices, defibrillators, ECG equipment, and pacemakers. Moreover, when in sole charge of a patient at night and on the weekend, details on ECMO (extracorporeal membrane oxygenation), dialysis, and Berlin heart are paramount. Knowledge about resuscitation and the handling of drugs that this involves, as well as about the equipment on the emergency trolley, is equally essential.

Therefore, as well as a firm grasp of diagnostic and therapeutic concepts on which this book intends to make a start, the practical on-site introduction of the new employee to all equipment and logistical processes is particularly vital. Recommendations here include the issue of an equipment operator's license, the restocking of emergency kits or emergency trolleys after deployment alongside the regularly practiced, independent use of equipment present at the patient's bedside—jointly with the nursing staff.

More important, however, is to also identify general prodromal signs of an impending emergency and thereby to prevent the situation from arising in the first place or to seek assistance. This entails that a minimum amount of information should be available on each patient in the intensive care unit (ICU), not only for a personal understanding but also for rapid responses to questions by any colleagues who are consulted. This textbook systematically describes the most important

details needed for the reliable understanding and communication of urgent and emergency situations arising in individual patients, but also those which are essential to understand in order to be able to anticipate the most common problems encountered in patients on a pediatric ICU.

What Should Be Actively Known About Each Patient?

- Age and weight
- Disease and day of surgery, including clinical course of previous disease, where applicable
- Hemodynamics in terms of:
  - Normal serial circulation
  - PDA-dependent systemic/pulmonary perfusion, systemic-to-pulmonary arterial circulation (PDA = patent ductus arteriosus)
  - Glenn or TCPC circulation (total cavopulmonary connection)
  - Left or right ventricular obstruction
- Data on hemodynamics: Blood pressure (BP), central venous pressure (CVP), microcirculation (MC), lactate, SvO_2 (central venous saturation) etc. (see individual chapters), drains, respiratory and ventilatory status, renal function, laboratory data
- Major diseases other than cardiac:
  - Respiration
  - Kidney
  - Gastrointestinal tract
  - Neurology
- Particular aspects of the previous history (endocrinology, syndromal diseases, particular social aspects, etc.)

After reading the book, any member new to the pediatric cardiology team should have a sound grasp of the individual details that encompass the intensive care patient's overall situation. This conceptual understanding should equally allow a structured handover to the next shift and other staff involved in the patient's care. Furthermore, the defined structure ensures that key points are not lost or overlooked, for example, even for a novice on the ICU.

Structured Handover

To be able to identify a patient's problems as quickly as possible and without the loss of relevant reporting, a chart for the structured handover of information to the next shift is essential.

The details required for this are best kept in one's head or noted down at close proximity – the (electronic) record serves as a reference. The reporting regimen

described should enable information to be passed on in the form of a common thread within a short space of time:

- Disease(s)/preexisting condition(s)/prior medication
- Change in general health (GH): Better – worse – the same (during shifts)
- Circulatory parameters: Blood pressure, CVP, MC, SvO_2, urine output, cardiovascular drugs
- Rhythm and pacemaker (PM) with antiarrhythmic therapy
- Lung function with ventilation parameters: Pressures, FiO_2 (fraction of inspired oxygen), MV (minute volume), Vti (tidal volume), and type of ventilation
- Kidney with urine output, specific urine characteristics, and diuretic therapy
- Drains and specific bleeding characteristics
- Neurological status with vigilance, analgesia, sedatives, specific aspects
- Gastrointestinal tract/metabolism with nutrition/medication to act on intestinal motility/glucose metabolism
- Laboratory values, particularly troponin I (TNI), coagulation, infection markers, liver function tests, microbial data
- Important social aspects for the patient's care/further procedure

In addition:

- Type of surgery and exact procedure should be documented by the admitting staff (whenever possible: Surgeon's drawing(s)).
- Ultrasound and X-ray findings should be entered in the chart by the surgeon and should be known.

To allow an efficient and well-structured handover, including to those who are less well versed, the following ground rules must be observed:

- Listeners listen acutely and are allowed to ask questions about individual points.
- Discussion of problems and establishment of the procedure, if possible, should be held jointly and comprehensibly for everyone at the end of the handover.

Part I

General Considerations on Pediatric Cardiac Intensive Care Medicine

O_2 Supply, CO_2, and Acid-Base Balance

1

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1.1 O_2 Supply and CO_2 Balance

1.1.1 O_2 Partial Pressure and Oxygen Cascade

In ambient air (and under standard conditions), an O_2 partial pressure (PO_2) of about 160 mmHg prevails.

Formula 1

$PO_2 = P_{atm} \times FiO_2$

At P_{atm} = 760 mmHg: 760 mmHg × 0.21 = *160 mmHg*

In the respiratory tract, inspired air is moistened (PH_2O = 47 mmHg) and then mixed in the ventilated alveoli with the CO_2 released. As a result, alveolar O_2 partial pressure (PAO_2) drops to about 100 mmHg.

PH_2O = hydrostatic pressure

Formula 2 = alveolar gas equation

$PAO_2 = (P_{atm} - PH_2O) \times FiO_2 - PaCO_2/RQ$

With an RQ (respiratory quotient) = 0.8 (mixed diet): (760 mmHg – 47 mmHg) × 0.21–40 mmHg / 0.8 = *100 mmHg*

Even in healthy subjects, however, arterial O_2 partial pressure (PaO_2) does not match alveolar partial pressure but is only about 95 mmHg (SaO_2 = 98–100%; see

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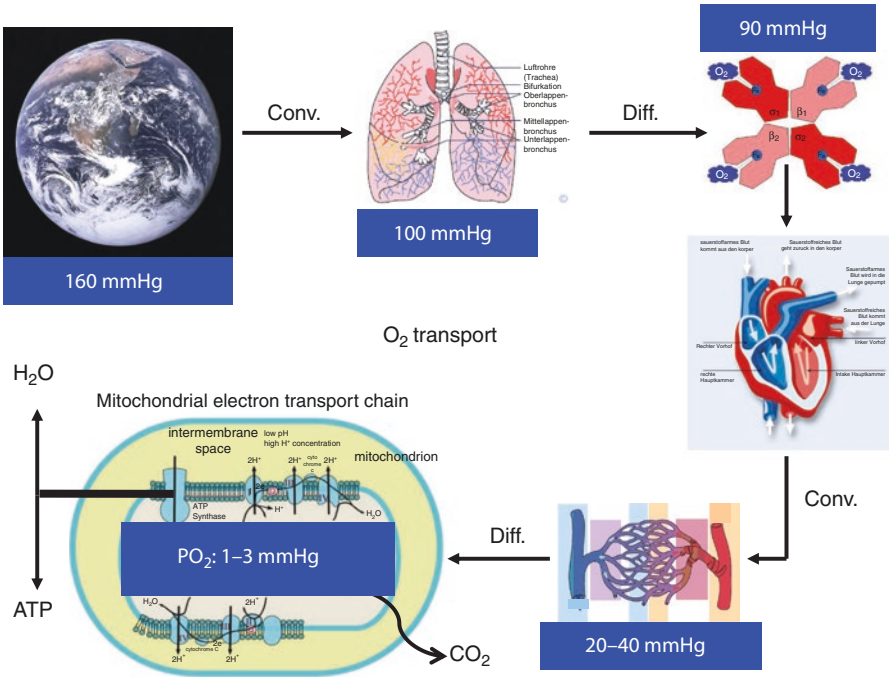

Fig. 1.1 O_2 transport from the atmosphere into the mitochondria. Conv. = convection, Diff. = diffusion

oxygen-binding curve, Sect. 1.1.3). This can be explained by diffusion losses (normally very small) and admixing of the "physiological shunt" (e.g., bronchial circulation, Thebesian = intracardiac veins) of about 1–3%.

As the diffusion distances from the capillaries to the cells are relatively large in tissue, the tissue O_2 partial pressure (PtO_2) falls to values of about 20–40 mmHg. Because of the difference in partial pressure between PaO_2 and PtO_2, the loaded oxygen is released by hemoglobin (Hb). During passage through the capillaries, the venous O_2 partial pressure (PvO_2) therefore approximates to the corresponding PtO_2 (i.e., PtO_2 and PvO_2 respond proportionately). Under normal circumstances, a PvO_2 of about 40 mmHg and venous saturation (SvO_2) of about 75% (normal arteriovenous SO_2 difference = about 25%) prevail in the veins. In the muscle, O_2 binds, for example, to myoglobin. Compared to Hb, it exhibits a left-shifted binding curve. In the mitochondria, ultimately there is only an O_2 partial pressure of 1–3 mmHg. The fact that oxygen is consumed in the mitochondria acts like a "gully" on the oxygen partial pressure gradients (Fig. 1.1)

Below What Value Does a Fall in PaO_2 Result in O_2 Deficiency (Dysoxia) of the Cells?

If PtO_2 falls below 20 mmHg, the diffusion distances in the tissue can no longer be sufficiently overcome, and oxidative energy production in the mitochondria ceases at a PtO_2 < 10 mmHg. The dysoxic threshold differs locally (i.e., from tissue to

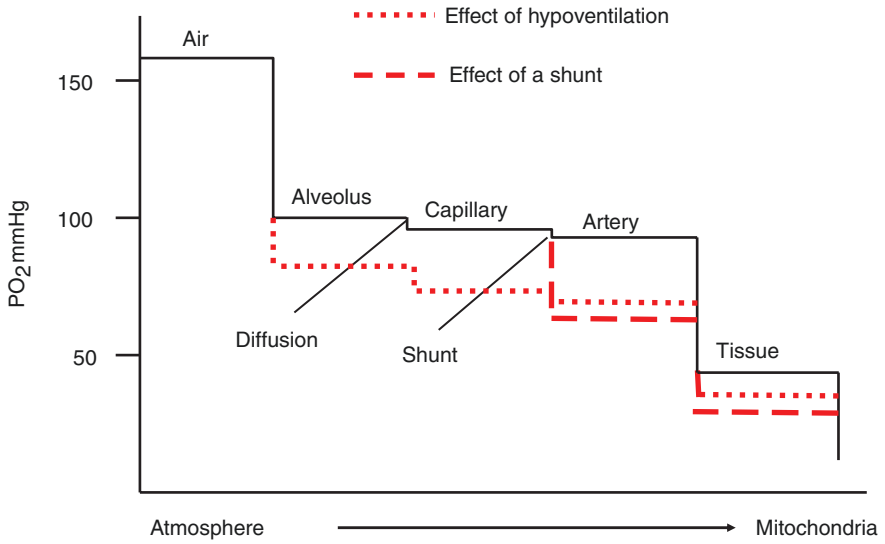

Fig. 1.2 Oxygen cascade

Table 1.1 Target values for preventing oxygen deficiency

	Noncyanotic	Cyanotic
PaO_2	> 60 mmHg (SaO_2 > 88%)	> 40 mmHg (SaO_2 > 75%)
PvO_2	> 30 mmHg (SvO_2 > 60%)	> 25 mmHg (SvO_2 > 40%)

tissue and from cell to cell) but is reported as a PaO_2 of <40 mmHg (SaO_2 < 75%) or a PvO_2 of 20–25 mmHg (SvO_2 = 35–40%) (Fig. 1.2).

If the drop in PaO_2 is too dramatic, the peripheral diffusion distances can no longer be overcome and the dysoxic threshold is reached.

The above remarks set important target values for intensive care medicine that need to be observed if oxygen deficiency is to be avoided (see Table 1.1).

Caution The stated target values for O_2 partial pressure must always be checked individually to ensure they are sufficient. Where necessary, higher ones can be targeted or lower ones tolerated. It should always be remembered that there is no room for maneuver at the lower limit and that therefore an alternative strategy for oxygenation (e.g., ECMO) must be available in the event of deterioration.

1.1.2 Causes of Reduced Oxygen Tension/Saturation in the Blood (PaO_2 or SaO_2)

Right/Left Shunt (= R/L Shunt)

When blood passes through a capillary bed (e.g., lung or peripheral tissue) without engaging in gas exchange, this is referred to as a shunt. In the case of an R/L shunt, part of the venous return flows past the lung and mixes with the arterial systemic

blood accompanied by a low O_2 partial pressure (= PvO_2). As a result, PaO_2 and SaO_2 are lower in the aorta (see saturation curve, Sect. 1.1.3). In the following, the saturations are discussed as surrogates for O_2 partial pressures.

The reduction in arterial saturation by a shunt depends directly on the extent of the shunt fraction (as a percentage of CO = cardiac output) and the level of SvO_2.

Extrapulmonary R/L Shunt

Example of Glenn anastomosis (= connection of the superior vena cava to the pulmonary arterial vascular bed as the first step to palliation in univentricular circulation):

In this case, ideally about 50% of the venous return flows via the superior vena cava into the lung and, thus oxygenated, reaches the ventricle via the pulmonary veins (pulmonary venous saturation [$SpvO_2$] ideally = 99%). The resultant pulmonary perfusion is solely responsible for oxygenating the patient. The remaining 50% of the venous return (= blood from the lower half of the body) is transported in a nonoxygenated form to the ventricle (SvO_2 = 50%) via the inferior vena cava. The ventricle serves as a mixing chamber, and the resultant SaO_2 in the aorta is approx. 75% (0.5 × 50% + 0.5 × 99% = 75%).

The ratio of pulmonary (Qp) and systemic perfusion (Qs) can be estimated from the saturation levels.

For parallel circulation (for further explanation please see Sect.15.11):

Formula 3

$CO = Qp + Qs$

Formula 4

$Qp/Qs = (SaO_2 - SvO_2)/(99 - SaO_2)$

Qp = pulmonary blood flow
Qs = systemic blood flow
99 = ideal pulmonary venous saturation ($SpvO_2$)

(This corresponds to the ratio of the arteriovenous saturation difference of the systemic circulation to the venoarterial saturation difference of the pulmonary circulation.)

In parallel circulation, a Qp/Qs ratio of 1.5:1 is the most suitable, since as a result, the volume load of the available ventricle is sustainably reduced, while at the same time, an adequate oxygen supply can still be delivered to the peripheral tissue. Such a Qp/Qs ratio is normally indicated by an SaO_2 of 75–85% (assuming there is no oxygen disorder of the lung and SvO_2 is >40%).

The calculation is of relevance, for example, in the following situations:

- HLHS (hypoplastic left heart syndrome)
- Following Norwood surgery or systemic-to-pulmonary artery shunt
- In ductal-dependent pulmonary perfusion

Caution An ideal pulmonary venous saturation ($SpvO_2$) of 99% is used in the calculation and not the actually existing $SpvO_2$ (measurement usually not possible). False low results may be obtained in oxygenation disorders of the lung (e.g., atelectasis, pneumonia) with reduced $SpvO_2$ (e.g., = 90%) (e.g., estimated, 75–50 / *99*–75 = 1.0; actual, 75–50 / *90*–75 = 1.6. Further examples in the specific part of the book).

Intrapulmonary R/L Shunt

An intrapulmonary shunt can be the cause of severe hypoxia, as, for example, in acute respiratory distress syndrome (ARDS). It occurs if alveoli are no longer ventilated as a result of compression or filling (edema fluid, secretions, cell debris). The ratio of ventilation (V) and perfusion (Q) is then equal to zero (V/Q = 0). As distinct from low V/Q areas, which are described as a "functional shunt" (see below), this is referred to as a "true shunt" (i.e., the blood flowing here is not oxygenated at all). Theoretically, a "true" and a "functional" shunt can be distinguished by an increase in inspired O_2 to 100%. With a "true" shunt, SaO_2 increases little if at all (see iso-shunt diagram according to Nunn, Fig. 1.3). By contrast, an increase in SaO_2 is to be expected when the proportion of low V/Q areas is high, as their PAO_2 increases despite low ventilation at an inspired O_2 of 100%. In practice, however, there is usually no clear separation, with the two forms of venous mixing usually present to differing degrees.

In addition, factors such as hypoxic pulmonary vasoconstriction (HPV, see below) affect the outcome. The pulmonary blood flow is redistributed by HPV. Poorly ventilated alveoli (= low PAO_2) are less perfused (vasoconstriction), and alveoli with a

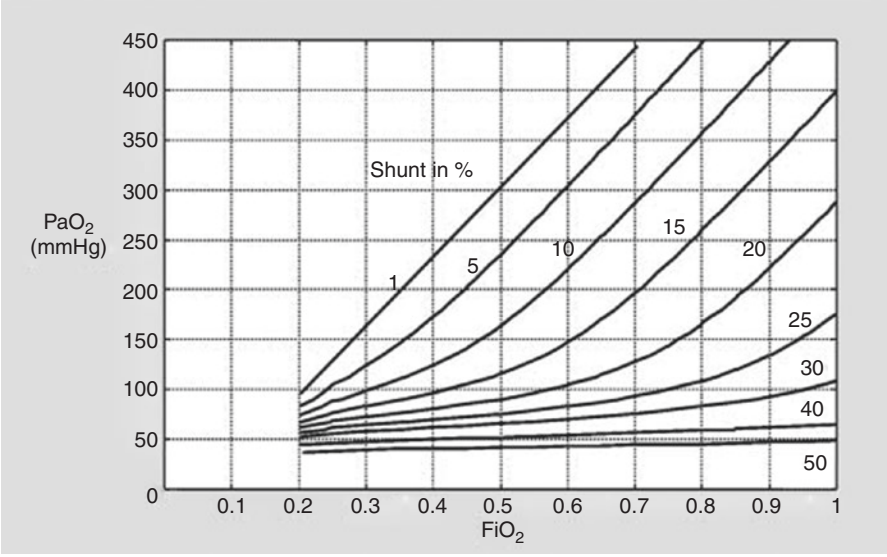

Fig. 1.3 Iso-Shunt-diagram (according to Nunn)