

Preoperative Medical Treatment of Congenital Heart Disease

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I. Congestive Heart Failure (CHF)

Definition

A clinical syndrome in which the heart is unable to pump enough blood to the body to its needs, to dispose of venous return adequately, or a combination of the two.

Etiology

.volume overload, pressure overload, and myocardial insufficiency
 .time of onset varies:

Table. 1
 Causes of CHF from Congenital Heart Disease

Age of onset	Cause
At birth	HLHS Volume overload lesions ; severe tricuspid or pulmonary insufficiency large systemic AV fistula
First week	TGA PDA in small premature infants HLHS TAPVR, especially pul. venous obstruction Critical AS, PS
1 - 4 wk	COA with associated lesion Critical AS large VSD, PDA in premature infants
4 - 6 wk	left to right lesions
6 wk - 4mo	large VSD, PDA anomalous left coronary a from PA

Clinical manifestations

Symptom ;

infants : poor feeding, tachypnea that worsens during feeding,
 poor weight gain, weak crying, cold sweat
 older children : shortness of breath (dyspnea) with activities

Sign ;

tachycardia, cardiomegaly, hepatomegaly, puffy eyelids

Adaptive mechanisms

- ① ventricular dilatation
 - ↑ end-diastolic volume → ↑ S.V.
 - but wall tension ↑ → ↑ myocardial O₂ demands
- ② ventricular hypertrophy;
 - pressure overload → heart wall을 thickening 시킴.
 - end stage에 가면 dilate
 - subendocardial region : underperfusion 되기 쉬움.
 - contractile state : normal과 failing myocardium의 중간상태
- ③ Adrenergic mechanisms
 - increased plasma concentration and urinary excretion of NE, Epi
 - ↑ β-adrenergic function : ↑ in myocardial contractility and heart rate.
 - ↑ α-adrenergic function : redistribution of cardiac output
 - ↑ to heart, CNS
 - ↓ to kidney, G-I, skin
- ④ Regional circulations
 - pulmonary dynamics : interstitial edema(tachypnea) → alveolar edema(rale, wheeze)
 - renal mechanisms :
 - ⓐ constriction of efferent glomerular arteriole and increased renal vascular resistance
 - greater fraction of plasma to be filtered
 - ↓ postglomerular hydrostatic pressure and ↑ postglomerular oncotic pressure
 - ↑ peritubular capillary uptake of proximal tubular fluid (Na)
 - ⓑ renin release
 - angiotensin I → angiotensin II → aldosterone → Na reabsorption ↑
- ⑤ Erythrocyte oxygen transport
 - rt shift of oxyhemoglobin dissociation curve via 2,3-DPG
 - ↑ oxygen unloading capacity
- ⑥ Atrial natriuretic factor (ANF)
 - role in volume and pressure regulation

Treatment

- 1. contents :
 - elimination of underlying cause
 - elimination of precipitating causes (infection, fever, anemia)
 - control of heart failure state
- 2. general measures;
 - oxygen(40-50%) with humidity
 - sedation : morphine or phenobarbital (occasionally, 1-2일간)
 - daily weight measurement
 - salt restriction : not indicated in infants.
 - older children (<0.5g/day)

- * monitoring for toxicity :
 - ECG
 - serum digoxin levels : therapeutic range 0.8-2.0 ng/ml
 - sampling : digitalization 후 3-5 일 지나서
6hrs after last dose or just before
a scheduled dose
- * predisposing factors to toxicity
 - ① high serum level
 - drug interaction (Quinidine, verapamil, amiodarone)
 - decreased renal excretion : premature baby, renal ds
 - ② increased myocardial sensitivity
 - myocarditis, myocardial ischemia
 - electrolyte imbalance (hypokalemia, hypercalcemia)
 - alkalosis, hypoxia
- * therapy for digitatis toxicity
 - ① discontinuation, ECG monitoring
 - ② PVC or supraventricular arrhythmia;
 - potassium chloride 3-5g/D in fruit juice
 - 0.5mEq/Kg/hr IV (max. 3mEq/kg/D)
 - Clix in advanced heart block, oliguria
 - ③ tachyarrhythmia;
 - lidocaine 1mg/kg bolus - 1-3mg/kg/hr(max. 5mg/kg)
 - dilantin 3-5mg/kg IV repeat 10-15min(max. 500mg/4hrs)
 - propranolol 0.01mg/kg IV per 2min (max. 0.1mg/kg)- 1-4mg/kg/D
 - P.O
 - DC version ; start at 0.5 joules/kg
 - ④ heart block;
 - Atropine : 0.01-0.03mg/kg per 4-6hrs (max. 0.4mg/kg)
 - transvenous pacing
 - ⑤ digibind(digoxin immune fab) : in accidental overdose
 - weight 20kg : 40mg (1 $\text{\textcircled{V}}$) IV for 30 min
- 3) Other inotropics

Epinephrine (Adrenalin)	IV 0.1 -1.0 ug/Kg/min
Isoproterenol (Isuprel)	IV 0.1 - 0.5 ug/Kg/min
Dobutamine (Dobutrex)	IV 5 -8 ug/Kg/min
Dopamine (Intropin)	IV 5 - 10 ug/Kg/min

 - @ dose related cardiovascular effects of dopamine
 - Renal vasodilation : 2-5
 - Inotropic : 5-8
 - mild vasoconstriction : > 10
 - Vasoconstriction : 15 -20
- 4) Afterload-reducing agents
 - ix : cardiomyopathy, postop, status, severe AR or MR
 - ① arteriolar vasodilators : propranolol과 같이 사용
 - hydralazine : (IV) 0.1-0.2mg/kg per 4-6hrs(max. 2mg/kg/dose)
 - (O) 0.75-3.0 mg/kg/D #2 or 4(max. 200mg/D)
 - ② venodilators;

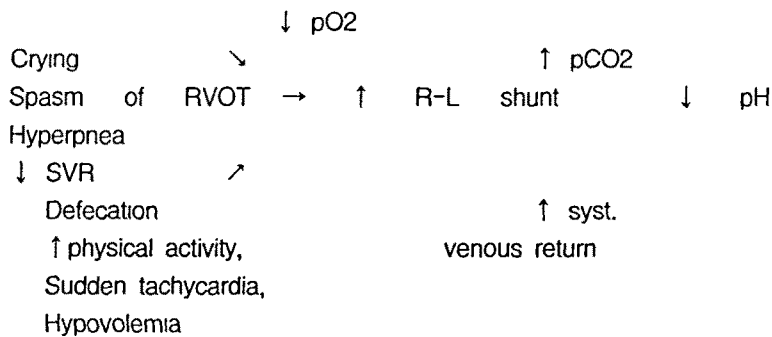
- nitroglycerin : (IV) 0.5-2.0 μ g/kg/min
- ③ balanced vasodilators; (postop rise in PA pressure에 사용)
 - captopril : (O) 0.5-6.0mg/kg/d. #1-4
12.5mg, 1-2번 in child
 - enalapril : (O) 0.1mg/kg, 1-2번/D
 - nitroprusside : (IV) 0.5-8 μ g/kg/min
 - prazosine(minipress): (O) 5 μ g/kg \rightarrow 25-150 μ g/kg/D. #4

II. Cyanosis

Definition :

a bluish discoloration of skin and mucous membrane resulting from an increased concentration of reduced hemoglobin (5g/100ml)
central cyanosis vs peripheral cyanosis

- 1) mechanism : vicious cycle



- 2) symptom & sign :

hyperpnea (rapid and deep respiration)
worsening cyanosis
disappearance of heart murmur
CNS symptom
death

- 3) Treatment : breaking cycle

- ① knee-chest position : ↓ systemic venous return (trap in legs)
↑ SVR by reducing arterial blood flow
 - ② morphine sulfate (0.2 mg/kg S. C or IM) : suppress resp. center
 - ③ oxygen
 - ④ NaHCO₃ (1mEq/kg IV) : reduce respiratory center-stimulating effect of acidosis
- ** ①-④로 full response 없으면 다음 Tx 실시.
- ⑤ Neo-syneprine (0.02mg/kg IV) : vasoconstrictor
 - ⑥ ketamine (1-3mg/kg IV for 1min) : ↑ SVR, sedation
 - ⑦ propranolol (0.01 - 0.25 mg/kg IV slowly): ↓ Heart rate

⑧ prevention : oral propranolol 2-6mg/kg/D #3 or 4

- * ductus dependent CHDs에서 ductus patency를 유지하기 위해 사용.
- * Indication
 - 1) source of pulmonary blood flow
TGA, pulm. atresia, severe TOF, Tricuspid atresia, severe ps
 - 2) source of descending aortic flow :
COA, hypoplastic left heart, IAA
- * use
 - PGE1(동아제약) : 0.05 μ g/kg/min로 시작, response에 따라 증감량
 - Eglandin(녹십자) : 5ng/kg/min로 시작.
response : PaO₂ 증가, cyanosis 호전
caution : light shielding
- * side effect :
apnea, hypotension, tachycardia, fever, irritability

III. Intervention

- A. Balloon atrial septostomy (BAS)
 - Indication ;
inadequate arterial oxygen saturations despite PGE1
TA, PA with IVS, critical PS, TGA, HLHS
restrictive foramen ovale : diameter < 4mm
- B. Palliative pulmonary balloon valvuloplasty
 - TOF : for decreasing the requiring of B-T shunt and transannular patch of RVOT reconstruction.
balloon/annulus ratio : 1.3-1.5
- C. pulmonary valvotomy
 - PA with IVS
critical ps
radiofrequency catheter 혹은 wire 이용하여 balloon 시행
- D. Stenting of ductus arteriosus
 - HLHS ; before cardiac transplantation
- E. Definitive interventional techniques
 - ① closure : ASD, VSD, PDA, collaterals (coil)
 - ② stents : branch pulm. a. stenosis, pulm. vein stenosis
 - ③ valvuloplasty : PS, AS