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Editorial

Relationship of high altitude and congenital heart disease

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1. High altitude

High altitude typically refers to elevations over 2000 m/6360 ft, but no single value is an adequate definition for all patients. During the ascent to high altitude, barometer pressure declines exponentially, and in keeping with Dalton's law, the partial pressure of oxygen falls accordingly.

Pressure chambers and air travel serve as a poor option to study the effect of hypoxemia on human physiology. Most natural is the systemic effects of high altitude on the natives and persons visiting hilly areas from low lands.[1]

The basis of the effects of high altitude comes from studies performed on aviators, mountaineers and natives of high lands.

More than 140 million people worldwide live at more than 2500 m above the sea level and around 80 million live in Asia.

2. High altitude and physiological adaptation

Peripheral chemoreceptor afferent activity rises hyperbolically as hypoxia increases and there is a phenomenon of ventilatory acclimatization. Initially due to acute hypoxia, heart rate increases, along with myocardial contractility and cardiac output. Later on, cardiac output falls at rest and on exercise with decrease in left ventricular work but increase in right ventricular work and pulmonary pressures.[2] Coronary circulatory flow increases along with progressive changes in right ventricular function, rising pulmonary pressures and chronic right ventricular pressure overload.[3] Unacclimatized person at high altitude develops hypoxic pulmonary vasoconstriction and rapid ascent can lead to subacute/chronic mountain sickness (Monge's disease) and high altitude pulmonary edema. This original article focuses on these aspects at physiological level and also in various cardiorespiratory diseases with special focus on congenital heart disease (CHD).

3. High altitude and cardiovascular illness

Although high altitude has a negative effect on preexisting respiratory diseases, we will focus on cardiovascular illnesses. High altitude has an adverse effect on the person from low altitude with rapid ascent and also on the person with underlying coronary artery disease, congestive heart failure, arrhythmias, systemic hypertension, and respiratory illness.[4]

4. High altitude and congenital heart diseases

As the original article highlights, high altitude has been linked with high incidence of CHDs like patent ductus arteriosus (PDA) and atrial septal defect (ASD) and their progression.[5] Both PDA and ASD are suspected clinically and later confirmed on investigations, especially Echocardiography. Transthoracic Echocardiography has limitations in detecting all cases of ASD, as there is a possibility of underestimation of this lesion. In the Baltimore-Washington infant study (Ferencz C, Rubin JD, Am J Epidemiol. 1985 Jan;121(1):31–6.), ASD was found in 0.0317%, and in the New England Regional Infant Cardiac Program (Donald C. Fyler, Pediatrics 1980), it was 0.0073%. The prevalence of ASD at the three high altitudes sites in the study at Qinghai Province 1988 was 2.4%.[6] The prevalence of PDA was 0.0089% and 0.01381% in Baltimore-Washington & New England Study. It was 1.2% at high altitude in Qinghai Province, which is much higher than the two larger studies. Failure of lower oxygen tension to constrict the ductus leads to patency of ductus arteriosus while the presence of high pulmonary vascular

Table 1 – Percentage-wise specific echocardiographic diagnosis of CHD in Dharan.[11]

Diagnosis	Number	Percentage
Acyanotic heart disease	**58**	**69**
VSD	49	58.3
ASD	4	4.8
ECD	2	2.4
Dextrocardia	3	3.6
Cyanotic heart disease	**26**	**31**
TOF	11	13.1
TAPVC	3	3.6
TGA with VSD	1	1.2
Unspecified	11	13.1
Grand total	**84**	**100**

VSD: ventricular septal defect, ASD: atrial septal defect, ECD: endocardial cushion defect, TOF: tetralogy of Fallot, TAPVC: total anomalous pulmonary venous connection, TGA: transposition of great arteries.
Adapted from: Incidence of congenital heart disease in tertiary care hospital, Shah GS, Singh MK, Pandey TR; Shah GS, Singh MK, Pandey TR; Kathmandu University Medical Journal (2008), Vol. 6, No. 1, Issue 21, 33–36.

resistance and right atrial pressure at high altitude inhibits early closure of foremen ovale. With physical development of the child and stretching of fossa ovalis along with incompetence of flap, ASD is established. Analogy can be drawn from high prevalence of ASD in TOF as the right ventricular pressure is high and right ventricular compliance is low from birth (JACC, 1988). At high altitude, these two anomalies are due to hypoxemia-induced failure of normal neonatal processes. Even the type of PDA at highland as compared to lowland is different, with larger ductal diameter, Type A lesions and high pulmonary arterial pressure (Bialkowski, 2003)[7] being more challenging for catheter closure.

The prevalence of CHD in India varies from 2.25 to 5.2/1000 live births.[8]

High altitude gives insight into the pathophysiology of both cyanotic and acyanotic heart disease in an interesting way. The patients with cyanotic CHD have more blunted hypoxic ventilatory response, which develops as early as 7–8 years, while the most blunted ventilatory response is seen in patients with maximum desaturation, which is corrected

Table 2 – Percentage-wise, sex-wise and age-wise specific echocardiographic diagnosis of CHD in Srinagar, data from Skims, Srinagar.[12]

CHD type	≤1 month		2 months to 1 year		2–5 years		6–12 years		>12 years		Total		Grand total (%)
	Male	Female	Male	Female	Male	Female	Male	Female	Male	Female	Male (%)	Female (%)	
Cyanotic													
TOF	10	2	13	10	4	5	2	1			30	18	48 (48)
TGV	10	1	5	7		2	1		1		17	10	27 (27)
Single ventricle	2		4	1							6	1	7 (7)
TAPVC	1		2	1	2						5	1	6 (6)
Tricuspid atresia	1	1									1	1	2 (2)
DORV	2	2	2	1		1	1				5	4	9 (9)
Truncus arteriosus									1		1		1 (1)
Total cyanotic CHD	26	6	26	20	6	8	4	1	2		65 (7.5)	35 (4.0)	100 (100.0)
Acyanotic													
ASD	31	29	37	40	17	16		2	1	3	86	90	176 (22.9)
VSD	20	25	64	66	29	19	7	7	1	3	121	120	241 (31.4)
VSD + (PDA/ASD/PFO)		2	16	4			1				17	6	23 (2.9)
PDA	47	39	30	34	14	12	1	6		1	92	92	184 (23.9)
AV canal defect	4	5	7	12	1	2	1				13	19	32 (4.1)
PS	7	3	15	13	7	6	3	1	1		33	23	56 (7.3)
Bicuspid aortic valve	1		1			1		1			2	2	4 (0.5)
Dextrocardia	2		2		1						5		5 (0.6)
Cardiomyopathy	1		7	9	4	7	1	2			13	18	31 (4.0)
Aortic stenosis	1		1		1			3			3	3	6 (0.7)
MVP MR	2			3	1	1	1				4	4	8 (1.0)
Coarctation of aorta									1		1		1 (0.1)
Total	116	103	180	181	75	64	15	22	4	7	390 (45.0)	377 (43.5)	767 (88.6)
Grand total	142	109	206	201	81	72	19	23	6	7	455 (52.4)	412 (47.5)	867

CHD: congenital heart disease, TOF: tetralogy of Fallot, TAPVC: total anomalous pulmonary venous connection, DORV: double outlet right ventricle, ASD: atrial septal defect, VSD: ventricular septal defect, PDA: patent ductus arteriosus, PFO: patent foramen ovale, AV: atrioventricular, PS: pulmonary stenosis, MVP: mitral valve prolapse.
Adapted from: Khurshid Ahmed Wanni, Naveed Shahzad, Prevalence and spectrum of congenital heart diseases in children; http://www.heartindia.net/article.asp?issn=2321-449x;year=2014;volume=2;issue=3;spage=76;epage=79;aulast=Wanni.