WWW.IJRPR.COM

International Journal of Research Publication and Reviews

Journal homepage: www.ijrpr.com ISSN 2582-7421

Case Report: -Management and Outcome the Diagnosis as a G3 A1 with GA with Previous FTND with RHD with MS with Mild Arotic Regurgitation with Mild Pyruvate Dehydrogenase Deficiency

[1]Ms. Bhawana Dhanvij, Mrs. Indu Alwadkar[2]

[1]Nursing Tutor, Florence Nightingale Training College of Nursing, Datta Meghe Institute of Medical Sciences (DU) Sawangi (M) Wardha, dhanvijbhawna@gmail.com, 8378025836

[2]Principal, Florence Nightingale Training College of Nursing, Datta Meghe Institute of Medical Sciences (DU) Sawangi (M) Wardha, 9960278995

ABSTRACT:

Regurgitation of the aorta On the other hand, congenital bicuspid aortic valve or aortopathy is more frequently associated with rheumatic heart disease than aortic regurgitation (AR) Because of the reduction in blood pressure and vascular system resistance that occurs during pregnancy, MR and AR are both easily tolerated, even when severe. The risk factors for maternal and perinatal morbidity and mortality include a higher risk when RHD during pregnancy (RHD-P). RHD, a rheumatic fever aftereffect that can damage the heart valves if left untreated, is twice as common in women. A genetic mitochondrial condition PDC) insufficiency is commonly followed by lactic acidosis, deteriorating neuromuscular and neurological aging, and typically, childhood death. A genetic metabolic mistake known as pyruvate dehydrogenase deficiency causes early infant death and long-lasting brain effects in survivors. Currently, prenatal diagnosis relies on fetal cell isolation for ensuing genetic and/or biochemical research. Pregnancy-related RHD (RHD-P) is linked to a higher risk of morbidity and death among pregnant women and newborns. RHD, a side effect of rheumatic fever that, if untreated, can harm the heart valves, is twice as common in women. This commentary gives context for prevention and therapy by giving a historical background. While knowledge, technology, economics, and social structures have led to vast variations in health systems throughout time and around the world, four fundamental themes underlie a large portion based on research on RHD during pregnancy (RHD-P): an RHD life-course approach for girls and women; the connection between RHD and poverty and inequality; calls for integrated care approaches to maximize maternal and perinatal outcomes; understanding of the usually complex care needs throughout pregnancy.

Keywords: Rheumatoid Heart Disease, Aortic Regurgitation, Pyruvate Dehydrogenase Deficiency

INTRODUCTION:

Rheumatic heart disease (RHD) is the chronic outcome of an aberrant immunological response to Streptococcus pyogenes, generally known as group A streptococcus (GAS), which results in scarring and dysfunction of the heart valves. Despite formerly being a serious public health issue in Europe and the US, the disease is still the main cause of death, heart failure, and stroke among young and middle-aged individuals in developing nations. Although accurate statistics are still lacking, it is suspected that at least 16 million people globally are affected by the condition, which also accounts for an estimated 300,000 premature deaths annually. Despite this, RHD has received very little attention from both researchers and funders in comparison to its overall impact. As a result, understanding pathophysiology has made only modest progress, which has complicated efforts to control disease and produce cutting-edge treatments and a potent vaccine.[1]

Both aortic regurgitation and mitral stenosis cause a decreasing pressure gradient to drive passive ventricular filling over a tiny aperture. Aortic regurgitation can be measured by Utilising Doppler ultrasound measurement of the velocity half-time, or time constant, mitral stenosis is successfully identified. 86 patients with varied causes of aortic regurgitation underwent continuous wave Doppler testing before cardiac catheterization or urgent aortic valve replacement. While the Doppler velocity half-time was calculated as the amount of time required for the diastolic aortic regurgitation velocity profile to degrade by 29%, the catheterization pressure half-time was calculated as the amount of time required for transvalvular pressure to decline by 50%.[2]

Cerebellar ataxia has been the most noticeable and even the sole neurologic impairment in patients with partial deficits of pyruvate dehydrogenase. It is unclear how this generalized enzyme shortage, which affects numerous organs and has activity levels between 15 and 30 percent below normal, might cause clinical symptoms that are specific to a small region of the nervous system. To determine the impact of partial enzyme deficits on pyruvate oxidation, we first contrasted the typical rate of pyruvate oxidation with the typical activity of pyruvate dehydrogenase in diverse areas of animal brains. The findings

show that pyruvate dehydrogenase deficiencies that are not severe enough to restrict pyruvate oxidation in the other areas of the brain we examined may be a hindrance to pyruvate oxidation in a region of the anterior cerebellar vermis.[3-4]

The Because it The pyruvate dehydrogenase complex (PDHc), an intramitochondrial multienzyme system, is essential for aerobic glucose metabolism by oxidative decarboxylation. It catalyzes the conversion of pyruvate to acetyl-CoA. Lactic acidemia and neurological anomalies are caused by genetic PDHc deficiencies. The majority of the time, the E1 subunit, the complex's first catalytic component, appears to be the source of the problem. The case study concerns a 6-year-old boy from Portugal who has modest neurological involvement, low PDHc activity, and no E1 according to immunoblotting tests.[5]

Poverty is a situation that leads to rheumatic heart disease (RHD). Research and funding for the global control of rheumatic fever and RHD have drastically decreased as a result of the near eradication of rheumatic fever and RHD in the industrialized world and the advent of competing major global epidemics like AIDS, TB, and malaria.RHD, however, has reemerged as a significant global health concern within the last ten years. The World Heart Federation and the World Health Organisation Federation aspire to see a reduction of 25% in cardiovascular disease-related fatalities, including RHD, by the year 2025.[6]

Case Presentation:

At the age of a 25-year patient diagnosed with diagnosis as a g3 a1 with ga with previous find with rhd with ms with mild aortic regurgitation with mild pyruvate dehydrogenase deficiency amenorrhea since 8 months, complaints of breathlessness since 5-6 days dom: 6 years g3p1l1a1 p1l1- 5 yrs/male/find/2.5 kgs a1- 3 yrs back/1.5months/induced with pills/d&c not done 2 tt ing received g3- present pregnancy menstrual history:- lmp - 07/11/22 edd - 14/8/23 pog- 35+1 weeks pmc:3-4 days/28-30 day/ reg/ avg/ no clots/ no dysmenorrhea.

History- newly diagnosed case of severe ms, mild tr, severe pah with rhd 2-3 days back no h/o in/dm/tb/ba/thyroid/epilepsy no h/o blood transfusion/in orofer no h/o major surgical illness no h/o recent hospitalization in present pregnancy. habits-sleep appetite- adequate, bowel bladder- regular, examination:- pulse: 82/min | resp: 20/min | b.p.: 116/78 mm of hg mmhg, respiratory system: b/l clear no basal creptsnyha grade iv cardiovascular system: s1s2 heard mid-diastolic murmur +central nervous system: conscious oriented to time place and person abdominal examination: p/a- uterus 34 weeks size relaxed long lie cephalic fhs/+/r/140bpm musculoskeletal system: gynecological examination: p/v-os closed uneffaced cervix- posterior station high up step of operation: name of the operation: elective less indication: g3p1l1a1 with 35+3 weeks ga with prev ftvd with rhd with severe ms with moderate tr with moderate ar with mild as with moderate pah in labor informed valid, written consent taken and patient shifted to ot table informed valid, written consent taken and under all aseptic precaution patient induced with general anesthesia and position given. under all aseptic precaution cleaning painting ad draping done and level of anesthesia checked. a pfanenstial incision was given and the abdomen opened in layers till the rectus sheath rectus sheath opened by blunt dissection. muscle separated peritoneum opened by blunt dissection and Doyen's retractor introduced u.v fold identified and opened and Doyen's retractor advanced to push the bladder downwards. lower uterine segment well formed and a trans curvilinear incision is given. A male baby was delivered via vertex presentation, ppvx, on July 13, 2013, at 9:52 a.m. The baby cried as soon as the chord was clipped, and the placenta was delivered with all cotyledons and intact membranes. Vicryl no. 1 interlocking sutures were used to seal the uterus. blood loss minimal hemostasis achieved, mop and instrument count confirmed. paracolic gutters checked. peritoneum closed by Vicryl taking continuous sutures. rectus sheath closed with vicryl with continuous sutures skin closed with mattress sutures hemostasis achieved, dressing and vaginal toileting done patient withstood the procedure well and was shifted to the ward. b/l tl done by modified Pomeroy's method.

Baby condition: sex: male, cry: immediately after birth, weight: 2.2 kgs Apgar: 8/10 9/10 baby shifted to relatives side. zero doses of polio, hep B, and BCG given medicine call done on 11/7/23 ivo k/c/o rhd with severe ms with pah with mild ar & tr adv: to continue as per protocol decided by cardiologist cardiology call was done i.v.o opinion in management adv- plan for less with cardiac backup avoid volume overload and tachycardia continues diuretics and beta blockers sos BVM in case required risks and benefits explained to relatives cvts call ivo k/c/o rhd with severe ms with pah with mild ar & tr : adv: patient ill require double valve replacement with/without tv repair continue beta blockers and diuretics.

Emergency med call ivo potassium value of 5.6 adv: injection calcium gluconate in 10 ml ns over 10 mins if ecg changes + and potassium value >5- in gi drip cardiology call done on 13/7/23 on day 1 of less ivo rhd with severe ms with pah with mild ar & tr adv: tab met xl 25 mg od if hr>90 bpm tab editor 10 mg od if bp >110/70 mm hg cardiologist consultation done ivo heart rate 128 bpm:adv: tab ivabradine 5 mg stat

Treatment in the hospital: in ceftriaxone 1gm iv 12 hourly x 7 days,in amikacin 500mg iv 24 hourly x 7 days, in metro 100 cc iv 8 hourly x 7 days, in pan 40 mg iv 12 hourly x 7 days,in Entomol 100ml iv 12 hourly x 3 days,tab tax 200mg bd x 2 days,tab pan40mg bd x 2 days,tab metro 400mg tds x 2 days,tab tramadol 50mg bd x 2 days,tab metal 25mg od x 7 days, tab editor 10mg x 7 days, zona suppository p/r tds x 5 days ,nebulization with Dublin, nebulization with budecort

Foleys removed on day 5 urine passed check dress done on day 5 mattress sutures, wound healthy no discharge check dress done on day 8 mattress sutures, wound healthy

No discharge asr done on day 9 mattress sutures.

Discussion:

A significant rise in cardiac output occurs during pregnancy, peaking Due to hormonally induced increases in heart rate, blood volume, and red cell mass, preterm labour begins in the second trimester and continues throughout pregnancy. Hypertension and peripheral vascular resistance simultaneously decline as a result of circulating prostaglandins, pregnancy hormones, and the placenta's low-resistance vascular bed. Blood pressure and cardiac output both rise more during labor and delivery as a result of discomfort and uterine contractions. The immediate aftermath of birth results in an additional increase in cardiac output due to the empty, constricted uterus' relaxation of caval compression and autotransfusion. Two weeks following delivery, the majority of hemodynamic abnormalities associated with pregnancy go away.[7] In northern Australia, rheumatic heart disease (RHD) still poses a serious health risk to indigenous women who are of childbearing age. Uncertainty exists regarding the impact of RHD on maternal outcomes within the present clinical paradigm.[8] This study examines whether it is morally acceptable to continue giving antenatal Anti-D Ig to all RhD-negative women when maternal blood-based fetal RHD genotyping could help identify which women do not require this medication.[9]

The tricuspid valve has an asymmetrical structure, and because of the surrounding muscle, its annulus is stiffer. The tricuspid valves normally coapt at or just below the tricuspid annular plane (5–10 mm of surface contact during systole between the leaflets). The RV's tenting of the valve leaflets wall during RV dilatation results in the elliptical configuration of the RV becoming more planar. The coaptation also moves from edge to edge. There might not be any coaptation of the leaflets during systole if there is considerable tethering. [10]

The frequency of concomitant mitral regurgitation and other valve diseases, such as MVP, as detected by color Doppler echocardiography in a sizable consecutive referral sample covering all MS degrees is first reported in this study.[11]

A genetic metabolic mistake known as pyruvate dehydrogenase deficiency causes early infant death and long-lasting brain effects in survivors. Currently, prenatal diagnosis relies on fetal cell isolation for ensuing genetic and/or biochemical research. Pyruvate dehydrogenase deficiency has occasionally been diagnosed and treated postnatally using magnetic resonance imaging and magnetic resonance spectroscopy. We provide an example of how these non-invasive techniques can be used to diagnose this issue in utero.[12-13]

Conclusion:

Maternal cardiac outcomes are significantly impacted by RHD. The prevalence of complications is expected based on known risk factors, and maternal and fetal deaths are minimal with current care techniques. RHD and rheumatic fever thrive in unsanitary environments and areas with weak public health infrastructure. Preventative measures are affordable, accessible, and economical. To eradicate RHD, current and accurate data coming from big Registries, like the ROPAC register, are essential for guaranteeing political and monetary commitment on a national, regional, and municipal level.

A mitochondrial condition of carbohydrate oxidation called primary pyruvate dehydrogenase complex deficiency (PDCD) primarily affects the brain and causes a decrease in ATP synthesis and an energy deficit. A group of abnormal metabolism (increased plasma pyruvate, lactic acidemia, and/or metabolic acidosis), abnormal brain imaging (dysgenesis of the corpus callosum, Leigh syndrome), and neurologic symptoms (congenital microcephaly, hypotonia, epilepsy, and/or ataxia) characterize primary PDCD the majority of the time. Developmental delays are almost always present.

References:

1. Parks T, Mirabel MM, Kado J, Auckland K, Nowak J, Rautanen A, Mentzer AJ, Marijon E, Jouven X, Perman ML, Cua T. Association between a common immunoglobulin heavy chain allele and rheumatic heart disease risk in Oceania. Nature communications. 2017 May 11;8(1):14946.
2. Teague SM, Heinsimer JA, Anderson JL, Sublett K, Olson EG, Voyles WF, Thadani U. Quantification of aortic regurgitation utilizing continuous wave Doppler ultrasound. Journal of the American College of Cardiology. 1986 Sep 1;8(3):592-9.
3. Reynolds SF, Blass JP. A possible mechanism for selective cerebellar damage in partial pyruvate dehydrogenase deficiency. Neurology. 1976 Jul 1;26(7):625-.
4. Prasad C, Rupar T, Prasad AN. Pyruvate dehydrogenase deficiency and epilepsy. Brain and Development. 2011 Nov 1;33(10):856-65.
5. João Silva M, Pinheiro A, Eusébio F, Gaspar A, Tavares de Almeida I, Rivera I. Pyruvate dehydrogenase deficiency: identification of a novel mutation in the PDHA1 gene which responds to amino acid supplementation. European journal of pediatrics. 2009 Jan;168:17-22.
6. French KA, Poppas A. Rheumatic heart disease in pregnancy: global challenges and clear opportunities. Circulation. 2018 Feb 20;137(8):817-9.
7. Siu SC, Colman JM. Heart disease and pregnancy. Heart. 2001 Jun 1;85(6):710-5.
8. Cupido B, Zühlke L, Osman A, van Dyk D, Sliwa K. Managing rheumatic heart disease in pregnancy: a practical evidence-based multidisciplinary approach. Canadian Journal of Cardiology. 2021 Dec 1;37(12):2045-55.
9. Teague SM, Heinsimer JA, Anderson JL, Sublett K, Olson EG, Voyles WF, Thadani U. Quantification of aortic regurgitation utilizing continuous wave Doppler ultrasound. Journal of the American College of Cardiology. 1986 Sep 1;8(3):592-9.

10. Samstad SO, Hegrenaes L, Skjaerpe T, Hatle L. Half time of the diastolic aortoventricular pressure difference by continuous wave Doppler ultrasound: a measure of the severity of aortic regurgitation?. Heart. 1989 Apr 1;61(4):336-43.

11. Samstad SO, Hegrenaes L, Skjaerpe T, Hatle L. Half time of the diastolic aortoventricular pressure difference by continuous wave Doppler ultrasound: a measure of the severity of aortic regurgitation?. Heart. 1989 Apr 1;61(4):336-43.

12. Robinson JN, Norwitz ER, Mulkern R, Brown SA, Rybicki F, Tempany CM. Prenatal diagnosis of pyruvate dehydrogenase deficiency using magnetic resonance imaging. Prenatal Diagnosis: Published in Affiliation With the International Society for Prenatal Diagnosis. 2001 Dec;21(12):1053-6.

13. Prasad C, Rupar T, Prasad AN. Pyruvate dehydrogenase deficiency and epilepsy. Brain and Development. 2011 Nov 1;33(10):856-65.