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DIFFICULTY IN MANAGING EISENMENGER SYNDROME: ABOUT A CASE SEEN IN THE TOAMASINA PEDIATRICS DEPARTMENT

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SUMMARY

Introduction: Eisenmenger's syndrome is the ultimate stage of congenital heart disease with untreated left-right shunt. Unfortunately, these patients are not always followed up in specialized centres. This makes their management in daily paediatric practice very complex. **Justification:** The objective of this study is to raise awareness among family doctors of the need to detect and then refer all heart diseases to a specialized centre at an early stage. All this in order not to delay treatment and to avoid their evolution towards the Eisenmenger complex. **Observation:** Our case concerns a 4.5 year old boy, admitted to the Paediatrics Department for an oedematous syndrome, with no personal or family cardiovascular history or reported malformation in the family, but in his personal history, we can retain a cough associated with repeated cyanosis, which may be in favour of a heart problem. On cardiac ultrasound, we objected to a heart disease with an Eisenmenger complex type with an aorto-pulmonary window. The patient was treated symptomatically, showing a transient improvement in his clinical condition. The subsequent evolution was enamelled by a succession of complex complications with multivisceral disorders: renal,

pulmonary, hematological and cutaneous. After 16 days in hospital, the patient died following a pulmonary embolism. **Conclusion:** Our observation illustrates the difficulty of managing this disease. Collaboration with family physicians for early diagnosis of any heart disease, in order to quickly refer it to a specialized centre, affects the immediate prognosis and long-term survival of these patients. Once installed, the Eisenmenger complex will be managed in a specialized and multidisciplinary environment, including a palliative care unit that will ensure a better quality of life for the patient.

KEYWORDS: Congenital heart disease, cyanosis, Eisenmenger's syndrome, polyglobulia.

INTRODUCTION

Eisenmenger's syndrome is the ultimate stage in the evolution of congenital heart disease with untreated left-right shunt. Its prognosis is better than that of idiopathic pulmonary arterial hypertension (PAH). Unfortunately, these patients are not always followed up in specialized centres. In practice, this makes our daily paediatric care very complex.

JUSTICATIVE

The objective of this study is to sensitize family physicians to the early detection and referral of all heart diseases, particularly V.I.C., in order not to delay their management and avoid progression to the Eisenmenger complex.

OBSERVATION

A 4.5 year old boy was admitted to the Pediatric service of CHU ANALAKININININA Toamasina ward in July 2018 for an edematous syndrome that appeared 10 days ago.

He was the youngest of two siblings from non-inbreeding parents, the eldest was in apparent good health. He had good psychomotor development but there was a staturo-weight delay. His vaccinations were up to date according to the national calendar.

The onset of the disease began at the age of three years with repeated pneumopathy and cyanosis without affecting his general condition, having been treated symptomatically. The occurrence of respiratory symptoms and cyanosis is becoming more and more frequent with an impact on his general condition motivating his hospitalization.

The echocardiogram performed showed congenital heart disease of Eisenmenger complex type with aorto-pulmonary window Initially, he was then put on digoxin and furosemide, potassium gluconate and a salt-free diet with a favorable evolution.

Six months later he was hospitalized for a second time following the reappearance of coughs with cyanosis of the extremities and edema of the generalized limbs.

He was in poor general condition at the entrance, with a blood pressure of 80/40 mm hg, intense asthenia, polypnea, fever at 38°c, perioral cyanosis and extremities refractory to oxygen administration. In addition, there is a deformation of the watch glass nails.

Other clinical examinations showed hepatomegaly, jugular vein turgor, hepatojugular reflux and generalized edema. Cardiac auscultation showed a stethacoustic anomaly with a systolic murmur 4/6th at the mitral focus, on the pulmonary level, diffuse bronchoalveolar rales were observed.

On the skin level, we observed an acne. The rest of the somatic examination was without particularity. At 7 days of his hospitalization, we found an epistaxis.

Polyglobulia at 16.5 g/dl hemoglobin and hyperleukocytosis at 16,000 GB/mm3 were revealed by the blood count. The CRP was elevated to 50 mg/l. Creatinine levels were slightly increased to 109 micromol/L. There was no ionic disorder initially on the blood ionogram. Cardiomegaly with a cardiothoracic index of 0.7 and pneumonia were detected on cardiopulmonary radiography.

Oxygen therapy, flurosemide, digoxin, acetylsalicylic acid and antibiotic therapy temporarily stabilized his condition. Finally, he died in a painting of chest pain, hemoptysis with hemoptoic tail, probably suggesting a pulmonary embolism 16 days after his hospitalization.

DISCUSSION

Eisenmenger's syndrome is the most feared complication of congenital heart disease with left right shunt. It has become rare in industrialized countries due to optimal perinatal care and early shunt suppression.

The presence of circulating endothelial cells is a factor in poor prognosis. It is due to a progressive alteration of the pulmonary arteriolar bed by an increase in pulmonary flow

caused by the left-right shunt. Pulmonary vascular resistance gradually increases until it becomes higher than systemic resistance leading to shunt inversion.[1]

Congenital heart disease is the leading cause of pulmonary arterial hypertension (PAH) in children.[2]

The WHO definition of PAH is an average pulmonary arterial pressure (P) greater than 25mmHg. According to the VENICE classification in 2003, revised at DANA in 2008, on pulmonary hypertension whose causes are left or pulmonary heart disease. PAH associated with congenital heart disease is the result of an increase in pulmonary flow related to shunt between systemic circulation and pulmonary circulation. Unlike idiopathic PAH due to an increase in vascular resistance due to intrinsic damage to the pulmonary arteriolar bed, PAH complicating congenital heart disease has a better prognosis if the diagnosis is made in time. Hopkins found that the 3-year survival rate of a patient with idiopathic PAH is 35% while it is 77% in Eisenmenger's syndrome.[3] According to a study conducted at Royal Brompton Hospital (England), the median life expectancy of a patient with Eisenmenger syndrome is less than 20 years compared to the general population and 40 years if the underlying heart disease is complex.[4] One of the main elements explaining the better tolerance of PAH related to Eisenmenger syndrome is physiological with the persistence of a "fetal" type of right ventricle function that is always subjected to high pressure after birth due to the non-restrictive shunt. It therefore remains enlarged and relatively adapted to the pressure overload, which prevents right ventricular failure, the main cause of death in other PAHs.[5]

Each left-right shunt initially leads to a particular symptomatology, which, as it evolves towards the development of pulmonary arteriolar disease, leads to a clinical picture common to all malformations. Thus, at first, high lung flow leads to feeding difficulties in infants, staturo-weight stagnation and repeated bronchopulmonary infections. Dilated pulmonary arteries compress the bronchi and unsystematic atelectasis is common. It is at this point, in general, that the decision to close the shunt is made if the diagnosis is made. As pulmonary arteriolar disease progresses, the signs of heart congestion improve, as increased pulmonary resistance no longer allows for significant left-right shunt. Without operation, resistance continues to rise until a bidirectional or inverted shunt is obtained: this is the definition of Eisenmenger's syndrome where PVRs become greater than 10 Wood units or 800 dynes-s cm5-1. Patients then have stress dyspnea, central cyanosis and digital hippocratism, polyglobulia secondary to hypoxemia, and specific complications.

The first published clinical description of Eisenmenger's syndrome was made by Paul Wood[6] in 1958. He reports 17.5% reaction from Eisenmenger. Clinical signs reported are a pulmonary ejection click, followed by a short systolic breath and a palpable and slammed B_2 with possibly a pulmonary insufficiency breath and signs of tricuspid leakage. A predominant desaturation of the lower limbs is suggestive of arterial duct with PAH. Edema of the lower limbs only appears very late (in case of failure of the subpulmonary ventricle). Our patient is hospitalized for advanced oedematous syndrome.

The chest x-ray initially favours pulmonary vascular overload, then the peripheral vascular framework becomes thinner and the proximal pulmonary arteries dilate. The cardiothoracic ratio is usually normal except in ASD where the right ventricle (RV) is dilated.

Cardiac ultrasound reveals the initial lesion (easily when it is a large VIC, the Doppler flow then highlights a VIC called with equal pressure without gradient between the ventricles, but with a right-left shunt at the color Doppler). Ultrasound is used to measure systolic pressure (equal to aortic pressure in the case of non-restrictive shunt) and sometimes diastolic pulmonary pressure but does not measure pulmonary resistance.[7]

Cardiac catheterization is at high risk in patients already in the shunt inversion stage. It is only of interest to patients for whom the problem of a possible surgical intervention arises. Lung pressures and resistance can be measured in this case in the basal state and under pulmonary vasodilators (02 and NO). Lung reactivity then indicates the potentially reversible nature of pulmonary arterial disease. The injection of iodine is particularly dangerous (risk of hypotension, thrombosis, and collapse). For those patients in whom a possible reversibility of pulmonary vascular disease is discussed, a pulmonary biopsy is sometimes indicated.[8]

Complications of Eisenmenger's syndrome

1. Polyglobulia and hemostasis disorders

Polyglobulia is a "salutary" reaction to cyanosis to improve oxygen transport in these patients (9, 10). The treatment of this polyglobulia is traditionally based on bleeding, which must be of small volume (5 to 7 ml/kg). Attempts at treatment with Hydrea (hydroxyurea) can give good results in experienced hands.

Symptoms of hyperviscosity are: Headache, Muscle fatigue, Vertigo, Blurred vision or amaurosis, Paresthesia, Psychomotor slowdown and chest and abdominal pain.

2. Stroke (Stroke)

The major risk factor for stroke, in addition to atrial fibrillation, is the existence of microcytosis, which can lead to martial failure due to excessive bleeding.[11]

3. Brain Abscesses

The absence of a pulmonary filter allows bacteria to pass through the systemic circulation.

4. Renal Impairment

It is common to see high urea, sometimes nephrotic syndrome.

5. Ventricular Impairment

Long-term hypoxemia can alter the myocardium, with significant ventricular hypertrophy in the event of marked cyanosis, and the development of myocardial fibrosis.

6. Sudden Death

During our patient's hospitalization, we identified multiple complications: epistaxis, pneumonia, acne, hyperazotemia and polyglobulia.

Early preventive treatment of severe left-right shunts is therefore essential.

Once pulmonary arteriolar disease has been established, it is not possible to close the shunt because the right ventricle would face suprasystemic pulmonary arterial resistance. The use of prostacyclins by analogy with primitive PAHs is constraining and gives partial results. The use of endothelin inhibitors is being tested.[12,13]

NO and sildenafil (Viagra) have shown some clinical efficacy.[14] Oxygen therapy has no proven efficacy but it can provide comfort because some pulmonary arterial hypertensions remain reactive to oxygen administration. The treatment is therefore essentially that of complications.[15]

Cardiopulmonary transplantation[16] remains the only surgical alternative for these patients. Unfortunately, this transplantation is not accessible under our current conditions. The only possibility is only palliative treatment, hence the importance of setting up a palliative care centre.

Nevertheless, psychological support for the family and the child is very important in the accompaniment of the disease.

Therapeutically, we were limited to symptomatic and palliative treatments.

Recently, an echocardiographic evaluation has been conducted to assess the patient's prognosis and survival.[17,18]

SUGGESTION

In the imagination of fundamentalist cardiologists, Eisenmenger's syndrome constitutes the failure of medical and surgical management of congenital heart diseases, hence the need to promote research, since clinical research obviously remains the strong link in progress for patients, but it can only develop in a sustainable way if it is based on solid foundations.[19]

The involvement of the various learned societies concerned obviously plays an important role in the dissemination of information to the health professionals concerned. They constitute an essential space for communication and exchange.

Patient associations are daily partners of HTAP networks. They have an essential support to families and doctors who rely on them for all actions: psychosocial aspects, solidarity, therapeutic education, professional life, information and support.

Eisenmenger's syndrome is one example among others of the interest of the Rare Diseases Plan. The multidisciplinary nature of the labelled networks, the confrontation of the cultures of PAH specialists and congenital cardiologists and the peer-to-peer relationships established have dramatically accelerated progress in these patients. This requires a complete clinical examination at each contact with the doctor, with an emphasis on the cardiovascular system.

CONCLUSION

Our observation illustrates the difficulty of managing this disease. Collaboration with family physicians for early diagnosis of any heart disease, in order to quickly refer it to a specialized centre, affects the immediate prognosis and long-term survival of these patients. Once installed, the Eisenmenger complex will be managed in a specialized and multidisciplinary environment, including a palliative care unit that will ensure a better quality of life for the patient.

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