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The Blue Girl

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Memories of my early years of establishing cardiology service from scratch in Qatar are still fresh and clear in my mind. Some cases are hard to forget because of the interesting social and human elements surrounding them. One such case was that of an Egyptian girl whose fate brought her to Qatar over 30 years ago because of cyanosis.

In June 1980, our cardiology section in Rumailah Hospital was two years old. By that time, we had acquired electrocardiography, phonocardiography, and M-mode echocardiography machines. We did not have a proper catheterization laboratory but we managed to do right and left heart catheterization in a room in the coronary care unit (CCU), using a CCU monitor and a small portable fluoroscopy machine attached to a video recorder. We used a portable x-ray machine to record one image at a time on a film to clarify or document a lesion of a coronary vessel by injecting a contrast agent, then quickly shoot a picture. We may have had to repeat such shooting because it gave us better images than our primitive video recorder. With this technique we were able to make a reasonable diagnosis for most adult cases.

Pediatric cardiac cases were problematic for me then because I was only trained to be an adult cardiologist. I only spent two months rotation in pediatric cardiology during my cardiology fellowship training in the USA at the University of Oregon in Portland, Oregon. After we moved from Rumaillah Hospital to the new Hamad General Hospital in 1982, I had to do a few simple cardiac catheterizations on children without enthusiasm. Therefore, we used to send most Qatari pediatric patients abroad for diagnosis and therapy until Dr. Gordon Folger, a senior pediatric cardiologist joined us in 1984. Dr. Folger was a very experienced American pediatric cardiologist with excellent qualifications. He underwent pediatric cardiology training at Johns Hopkins Hospital under the supervision of the late Helen B. Taussig, a world-renowned pediatric cardiologist. Dr. Folger established the pediatric cardiology section at Hamad General Hospital. I used to call Dr. Folger "the old man" because he was the most senior member of our department. At first he protested, but later he accepted it with grace when he realized that such a title was a sign of respect in our society.

During the early 1980s, my friend Dr. Gelal El-Said, a well-known Egyptian cardiologist, used to give us priority on his trips outside Egypt. Dr. El-Said is a senior cardiologist with excellent knowledge of both adult and pediatric cardiology. He is a first class clinician and teacher. During his visits, we would present to him pediatric cases referred to us. If the number of pediatric cardiac cases were small for discussions, he used to go to the pediatric ward, talk to the pediatricians about patients with murmurs in their ward, and add new cases for our meetings. He was keen to teach.

In June 1980, during one of Dr. El-Said's visits to our department, the pediatricians presented an interesting patient during one of our cardiology conferences. The patient was a 14-year old Egyptian girl with cyanosis (i.e. blue lips and blue fingers). She was seen a couple of years earlier in Egypt by general practitioners who labeled her as a case of cyanotic heart disease. Her father, a university professor, had consulted a cardiac surgeon in Egypt. The surgeon recommended surgery for the little girl but the father could not afford the cost of surgery. He decided to seek employment in the Arabian Gulf to improve his income in order to save enough money for his daughter's surgery. He ended up teaching at the University of Qatar in Doha.

The patient was being followed up in the pediatric department in Rumailah Hospital for two years as a case of congenital cyanotic heart disease and recurrent pneumonia. During those two years she was hospitalized several times for cough and "pneumonia". On each of her several admissions, the radiologist reported her chest x-ray as "acute pneumonia". Her CBC revealed polycythemia. The

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pediatrician treated her with antibiotics on each hospitalization. Her cough resolved but not the x-ray abnormalities!

Evaluation in our department revealed that she did have dyspnea, cyanosis, and clubbing. She had a flow murmur at the left sternal border compatible with an innocent murmur. Her chest x-ray showed a large density over the right lower lung field (Fig. 1).

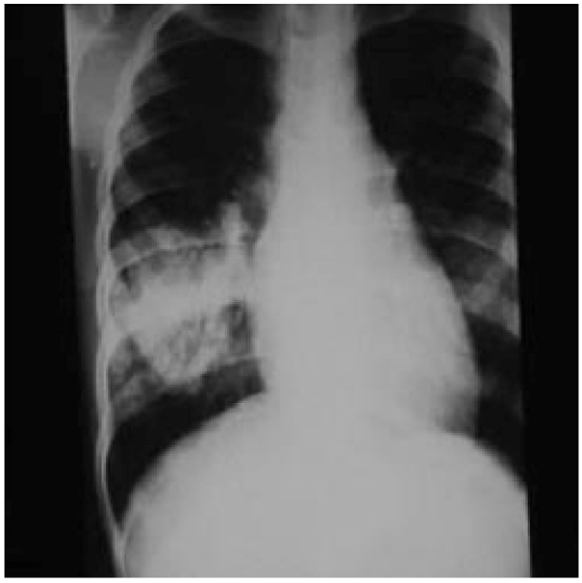

Figure 1. CXR shows a rounded, well-delineated shadow on the right lower lung zone.

The electrocardiogram was normal. The big blow to the diagnosis of cyanotic heart disease came from her echocardiography, which revealed a completely normal heart. *So why is the child cyanotic?* I wondered.

Dr. Galal El-Said examined the child. He found no significant abnormality on auscultation of the heart over the anterior chest wall but, he heard a loud murmur when he listened to the back corresponding to the shadow on the x-ray. He thought that the shadow on the CXR was not due to "pneumonia". "It must be an AV [arteriovenous] fistula", he said. He had seen a case like her before. He suggested that we perform right heart catheterization the following day to confirm the diagnosis.

Right heart catheterization is a simple procedure that can be done by a resident or an intern after a short period of training, but in Rumailah Hospital at that time, it was a sophisticated procedure because it had never been done before we established a cardiology section. It was new, not only in Doha but also in most Arabian Gulf States then. I remember in 1979, I received an urgent call from a health official in a Gulf state asking me if we had Swan-Ganz catheter in Qatar. It was needed by a visiting American cardiologist from Houston, Texas who was brought in to manage a very sick member of the royal family in that country where no cardiology service was established. I told them that we did have it. They suggested they would send an airplane that night to Qatar and a Boeing 747 airplane was sent urgently. I had to rush to Doha Airport at night with the catheter. The airport security took me in a car to the airplane, which stopped briefly on the runway for me to hand over the catheter to a health official. It then took off with the valuable catheter. The expensive airplane trip to pick up one relatively cheap catheter convinced our reluctant Rumailah Hospital administrators in Doha of the importance of such catheters. They were eager then to approve stocking up such catheters in our hospital only after that incident.

The following day, I prepared the girl for right heart catheterization on a wooden couch, which was made locally by our hospital carpenter for the catheterization laboratory according to my drawings and specifications. I remember telling the carpenter to avoid using nails except on the periphery of the couch. We used that couch for catheterization or pacemaker insertion because no radiolucent bed or catheterization table was available in our Rumailah Hospital then.

I sent for Dr. El-Said to join me before I punctured the vein. I needed his input for the final step. Finally, he entered the room, smiling, in a good mood, and rubbing his hands as if he were feeling cold.

"What are you waiting for?" he said. "You will see it; it will be nice."

"Why are you so excited Gelal?" I asked.

"This is an exciting case", he replied.

We passed a catheter from the basilic vein into the pulmonary artery through the right heart. We manipulated the catheter toward the area corresponding to the shadow on the chest x-ray film, while Dr. El-Said narrated his unsuccessful fishing trip the evening before. After five minutes of being in a boat at sea, he developed atrial fibrillation. "Every time I ride in a boat, I get atrial fibrillation." He interrupted his story when he saw the tip of the catheter in the right place. "Let us inject here", he said. We injected contrast. The AV malformations lighted up like falling stars in the middle of the night (Fig. 2). So, pulmonary arteriovenous malformations (PAVM) or pulmonary arteiovenous fistulae (PAVF) was confirmed. It consists of abnormal communications between the pulmonary veins and arteries. Such congenital malformation is rare. One study detected only 3 cases in 15,000 consecutive autopsies [1]. I had never seen one before. In 1939 a case diagnosed during life was published [2]. The first report of PAVM was described in 1897 [3]. It was an autopsy finding.

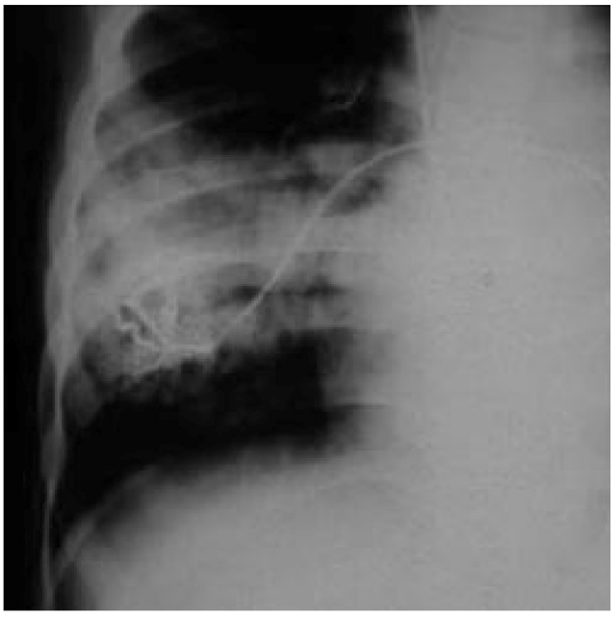

Figure 2. Contrast injection demonstrating at least three pulmonary A-V fistulae. (A-V = arteriovenous).

Simple right heart catheterization was adequate to prove that Dr. El-Said's diagnosis of AV fistula was correct.

I remember two other interesting right heart catheterizations we performed that year. One was a case of a 30-year-old obese female admitted with a two-day history of severe shortness of breath and whom I catheterized for diagnostic purposes. She had severe pulmonary hypertension and highly elevated right ventricular pressure. The procedure took only ten minutes and was quick smooth and uneventful. She probably had showers of pulmonary emboli. She died 12 hours after the procedure. Some physicians who were not familiar with such a simple procedure wondered if the patient died because of the Swan-Ganz catheter. The second case was one of our cardiology residents, Dr. Thomas. During morning rounds, while discussing a patient with a ventricular septal defect (VSD), he told me that he also had a VSD, diagnosed in India. He was asymptomatic but wished to know the shunt size. I told him we could get the answer immediately. He agreed to the procedure and he lay down on an empty CCU bed. I inserted a thermodilution catheter though the femoral vein, obtained saturations and cardiac output while our staff were in the room watching. The shunt was very small. We pulled the catheter out and he continued his CCU rounds.

At that time we had neither a cardiac nor chest surgeon in Qatar. The cyanotic Egyptian girl was lucky, not only because a definitive diagnosis had been made, but also because a few days after she underwent cardiac catheterization, a Lebanese chest surgeon, Dr. Saleem, from the American University of Beirut visited our hospital. I asked his opinion about the case. After he saw the X-ray and

catheterization film, he was excited about the case. "She should not be left alone with that huge malformation and such a severe degree of right to left shunt", he said. If left untreated, our young patient was at high risk of complications such as strokes, transient ischemic attacks, brain abscesses, and seizures.

Surgery in Qatar was the only option for her. In fact the literature twenty years ago tells us that surgery (ligation, local excision or lobectomy) was the only method of treatment any place in the world then. Embolization therapy using coils was in its infancy and was first reported in 1977, three years earlier than our case. Coil embolization therapy is now used routinely by the pediatric cardiologists and radiologists in our hospital, but at that time, we did not dream of such therapy in Qatar.

We put her on prophylactic intravenous antibiotics during surgery. She entered the operating theater blue like the Nile and came out pink like a flower. Her hospital course was smooth.

The following year the patient and her family returned to Egypt for good. Dr. Gelal El-Said saw the family a year later in Cairo. Dr. El-Said informed me, "The girl is well. She is pink and blushes normally."

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