

## Letters to the Editor

### Time for a Coronary Intervention Registry In Saudi Arabia

*To the Editor:* In an emerging coronary interventional practice, such as in Saudi Arabia, where there are limited centers, there is a great opportunity to establish a registry of coronary intervention, similar to the NHLBI (National Heart, Lung and Blood Institute) Registry for angioplasty and the NACI (New Approaches to Coronary Intervention) Registry for new devices of the United States.<sup>1</sup> Why should we do this? The proposed registry would provide us with a benchmark that would reflect current interventional practice in the Kingdom. Secondly, it would provide us with the power of adequate numbers of patients (the *n*) that would meet standards for publication in international meetings and journals. Alone, no center in the Kingdom has a volume that compares with busy centers in Europe and the USA. Further, the registry would serve as the steppingstone for planning and coordinating prospective trials.

The implementation of this registry will require cooperation among interventionists and the centers. The database standards have already been set and published by the American College of Cardiology and Society of Coronary Angiography and Intervention, and these could be adapted for local use.

A five-year search of the Medline in Cardiology, from 1994 to 1998 (Silver-Platter Inc.), reveals 370 citations from Saudi Arabia. It includes pioneering and major work from Saudi Arabia in the field of therapy for valvular heart disease and the congenital heart diseases. Yet there is not a single reference on coronary intervention. It is time to amend this.

**Waqar Habib Ahmed, MB, FACC**  
**Khaled F. Al-Shaibi, MB, FACC**  
King Fahad Armed Forces Hospital  
P.O. Box 8649  
Jeddah 21429, Saudi Arabia

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### Aplasia Cutis Congenita

*To the Editor:* We read with interest the article on aplasia cutis congenita by Bhatt et al. and would like to share our experience with an unusual case associated with dysmorphic facies, cranium bifidum occulta, anoph-

thalmia, dextrocardia and distal limb malformation. This association, to the best of our knowledge, has not been reported before in the Middle East.

A full-term male baby was born in the hospital by normal vaginal delivery to a 35-year-old Saudi, gravida 6 para 5, following an uncomplicated pregnancy. There was no history of obstetrical trauma. The parents were non-consanguineous. There was no family history of any congenital malformation. At birth, the baby's weight was 2750 g, length was 48 cm and head circumference was 34 cm. Physical examination revealed a 3x3 cm clover leaf-shaped sharply demarcated skin defect surrounded by a patch of alopecia in the scalp over the vertex in the midline (Figure 1). The floor of the defect was covered by a yellowish membrane formed of dura. Palpitation revealed underlying bony deficiency covering an area of 8x5 cm.

The face was slightly asymmetrical, with low-set and malformed ears, broad and bulbous nose, narrow palpebral fissures and absent eyeballs. The hands showed flexion contracture of the fingers, clinodactyly of the index fingers, postaxial polydactyly and hypoplastic nails. The feet showed bilateral rockerbottom deformity, fibular polydactyly and hallux valgus. The baby was found to have cranium bifidum and dextrocardia on investigations, including skiagraphy and CT scan. The baby died of respiratory infections within the first few days of life.

Aplasia cutis congenita (ACC) is a heterogenous group of disorders first reported by Cordon in 1767.<sup>2</sup> Although it is a sporadic disorder, both autosomal recessive and dominant patterns of inheritance have been observed. Frieden<sup>3</sup> classified ACC according to the number and location of lesions and the presence or absence of associated malformations into nine groups: 1) scalp ACC without multiple anomalies; 2) scalp ACC with associated limb anomalies; 3) scalp ACC with associated epidermal and organoid nevi; 4) ACC overlying embryologic malformation; 5) ACC with associated fetus papyraceous or placental infarct; 6) ACC associated with epidermolysis bullosa; 7) ACC localized to extremities without blistering; 8) ACC caused by teratogens; and 9) ACC associated with malformation syndromes.

Associated developmental defects are rare. They may be associated with some malformation syndromes, such as Patau's syndrome (trisomy 13), Wolf-Hirschhorn syndrome (4 p-), ectodermal dysplasia, Johanson-Blizzard syndrome, focal dermal hypoplasia, amniotic band disruption complex and gonadal dysgenesis.<sup>3</sup>

The clinical picture of our case resembles Patau's syndrome. We did not perform chromosomal studies due to lack of facilities. However, chromosomal studies are probably warranted in any child with scalp aplasia cutis congenita associated with multiple congenital anomalies when a specific syndrome is not identified, since mosaicism and partial trisomy 13 can show less severe

phenotypes. Once evaluation has been completed, and a specific diagnosis is established, genetic counselling of the family regarding the risk of recurrence can be accomplished.

**Suresh Dargan, MBBS, MS**  
Department of Orthopedics  
**Sandeep K. Dey, MBBS, MD**  
Department of Pediatrics  
Government General Hospital  
Rafha, North Zone  
Saudi Arabia

#### References

1. Bhat FA, Mathur BS, Hussain MT, Khalil F. Aplasia cutis congenita: a report of five cases in Riyadh. *Ann Saudi Med* 1998;18:185-6.
2. Cordon M. Extrait d'une lettre au sujet de trois enfants de la même mère nés avec partie des extrémités dénuée de peau. *J Med Chir Pharmacie* 1767;26:556-7.
3. Frieden IJ. Aplasia cutis congenita: a clinical review and proposal for classification. *J Am Acad Dermatol* 1986;14:646-60.

#### Reply

I read with interest the letter from Drs. Dargan and Dey, regarding our article on aplasia cutis congenita. The case they reported is a rare and unusual presentation of aplasia cutis congenita associated with various malformations. It obviously fits into group 9 of Frieden's classification. None of the cases reported by us had associated deformities and that makes their report worth publishing. I appreciate Drs. Dargan and Dey's mention of Frieden's Classification of ACC.

We recently had one more case of ACC of posterior scalp without any other anomaly. The defect was closed directly.

**Bhagwat S. Mathur, MCh, FACS**  
Plastic Surgeon  
Riyadh Medical Complex  
P.O. Box 7914  
Riyadh 11472, Saudi Arabia