

## REFERENCES

1. Kozlowski, Mazel. Asphyxiating thoracic dystrophy without respiratory disease. *Pediatr Radiol* 1976, 5: 30-33.
2. Jequier JC, Savreau—Ethier M, Gregoire H. Asphyxiating thoracic dysplasia. *In: Progress in Pediatric Radiology: Intrinsic Disease of the Bones.* Ed. Kaufman HJ. Basel, Karger, 1973, p 184.
3. Oberklaid F, Danks DM, Mayne V, Campbell P. Asphyxiating thoracic dysplasia: Clinical, radiological and pathological information on 10 patients. *Arch Dis Child* 1977, 52: 758-765.
4. Shah KJ. Renal lesions in Jeune's syndrome *Br J Radiol* 1980, 53: 432-436.
5. Lipson MJ, Waskey J/ Rice *et al.* Prenatal diagnosis of asphyxiating thoracic dystrophy. *Am J Med genet* 1984, 18: 273-277.

## Citrobacter Sepsis in Infants

**Kari Saraswathi**  
**Anuradha De**  
**Alka Gogate**  
**Armida R. Fernandes**

Gram negative bacilli are important agents causing neonatal sepsis(1,2). Infections may be acquired from the mother's genital tract or nosocomial infections acquired from the delivery room or from the nurseries(3). Multidrug resistant Gram negative bacilli like *Klebsiella*, *Citrobacter*, *Salmonella* sp. and *Pseudomonas* are gaining importance in the recent years(1-4). *Citrobacter*

species, recognized as saprophytes earlier are now known to cause neonatal meningitis, brain abscess, subacute bacterial endocarditis and sepsis(2,3). This communication presents clinical profile and antibiotic susceptibility pattern of five cases of *Citrobacter* sepsis encountered in the pediatric wards of our hospital between the period June 1992 to May 1993.

### Case Reports

Five cases of *Citrobacter freundii* were isolated from blood cultures of 2015 children (age group 0-1 year) with suspected sepsis, who were admitted in our hospital between June 1992 to May 1993. The total number of positive cultures were 480 (23.82%), out of which 5 were *Citrobacter freundii* (1.04%). Among the 5, two were neonates born in our hospital and 3 were outborn babies. Out of 5 cases, three were preterm neonates. The clinical signs and symptoms, complications, investigations and antibiotic sensitivity pattern of five infants suffering from *Citrobacter* sepsis are given in *Table I*. All the 5 *Citrobacter* sp. were resistant to ampicillin, gentamicin,

*From the Departments of Microbiology and Pediatrics, L.T.M. Medical College, Sion, Bombay 400 022.*

*Reprint requests: Dr. Anuradha De, Lecturer in Microbiology, LTMMC, Sion, Bombay 400 022.*

*Received for publication: March 12, 1994;*

*Accepted: November 4, 1994*

TABLE—Clinical Profile of Five Infants with *Citrobacter Sepsis*

Features	Patient number				
	1	2	3	4	5
Age at admission	1 day	15 days	2 months	1 year	1 day
Sex	Male	Female	Female	Female	Female
Gestational age	26 wks.	28 wks.	Not known	Not known	30 wks.
Birth weight (g)	800	1125	2500	Not known	1200
Symptoms and Signs					
Hypothermia	+	-	-	-	+
Fever	-	+	+	+	-
Lethargy and poor feeding	-	+	+	+	-
Pallor	+	+	+	+	+
Icterus	+	-	-	-	+
Ecchymoses	-	+	-	-	+
Convulsion	+	+	-	+	-
Diarrhea and vomiting	-	-	-	++	-
Abdominal distension	+	-	-	-	+
Respiratory distress	+	+	++	+	++
Unhealthy umbilical stump	+	+	-	-	+
Complications					
Sclerema	+	+	-	-	+
Abscesses	+	-	-	-	+
DIC	-	+	-	+	-
Hepatitis	+	-	-	-	+
Investigations					
Hb (g/dl)	10	12	11	7.5	8.5
Leucocytosis/leucopenia	+/-	+/-	-/-	-/-	-/-
Neutrophilia/neutropenia	-/-	-/-	-/+	-/+	-/+
I:T ratio	0.2	0.4	0.3	0.2	0.3
Toxic granulations	+	+	+	-	+
C-reactive protein	+	+	+	+	+
SGPT (IU)	60	Not done	Not done	Not done	45
VDRL test	NR	NR	R>1:64	NR	NR
Antibiotic sensitivity pattern					
Cefotaxime	R	R	R	S	S
Amikacin	R	S	S	R	R
Netilmycin	S	S	S	R	R
Outcome	Died	Died	Died	Died	Died

chloramphenicol, tetracycline and cotrimoxazole. Stool and cerebrospinal fluid (CSF) of these 5 infants were negative for *Citrobacter* sp. In 2 infants, *Citrobacter freundii* was isolated from the umbilical stump and in two cases from subcutaneous abscesses. This organism could not be isolated from environmental cultures. Cultures of the mother of the three preterm neonates was negative for *Citrobacter freundii*. Two preterm babies had raised SGPT. In one case VDRL was strongly reactive (R >1:64). All the five infants expired despite appropriate antibiotic therapy.

#### Discussion

In the recent years *Citrobacter* sp. is recognized to be a potential pathogen in neonates and in infants with a propensity for producing a necrotizing meningoencephalitis(5). Nosocomial outbreak of citrobacter due to umbilical colonization has also been reported(6). Multidrug resistant citrobacter sepsis in infants have also been reported, though source of infection and mode of transmission have not been defined(1,2). Recently there was a report of nosocomial neonatal septicemia due to *Citrobacter freundii* which was due to contamination of delivery room unit as a result of drainage pipe leakage(3).

All the investigations in the five infants gave a picture of sepsis. All the five cases reported by us showed respiratory distress and tachypnea. This is a warning signal for conventional organisms like Group B streptococci and other Gram negative bacilli, but when these are ruled out *Citrobacter* sp. must be kept in mind(3). Though *Citrobacter diversus* was isolated in an epidemic of nosocomial meningitis(6), in our study

only *Citrobacter freundii* was encountered. *Citrobacter* was isolated from the umbilical stump of two patients, but stool and CSF cultures were negative. The same organism could not be detected from environmental cultures. This organism was not isolated in any older children during the same period. All patients were in age group 0-1 year.

Complications seen were sclerema(5), disseminated intravascular coagulation (DIC) and hepatitis. Multiple abscesses were observed in two cases. Extreme prematurity is a predisposing factor for citrobacter sepsis(7), and three of our cases were premature.

Multidrug resistant strains do not always suggest them to be of nosocomial origin. Common source of infection could not be detected in our study. So it is not a nosocomial outbreak. Vertical transmission was excluded in the three neonates as the culture studies of their mothers were negative.

To conclude, *Citrobacter* sp. has become an important pathogen in the recent years in causing neonatal sepsis, apart from *E. coli*, *Klebsiella*, *Pseudomonas*, etc. It is important to know the source of infection as mortality is 100% in our study despite appropriate antibiotics and other supportive measures.

#### REFERENCES

1. Chaturvedi P, Agarwal M, Narang P. Analysis of blood culture isolates from neonates of a rural hospital. Indian Pediatr 1989, 26: 460-465.
2. Sugandhi RP, Beena VK, Shivananda PG, Baliaga M. Citrobacter sepsis in infants. Indian J Pediatr 1992, 59: 309-312.

3. Raghu Raman TS, Jayaprakash DG, Singh D, Krishnamurty L, Menon PK. Citrobacter septicemia in neonates. *Indian Pediatr* 1993, 30: 516-520.
4. Sashidharan CK, Rajagopal KC, Panicker Jayaram CK. *Salmonella typhimurium* epidemic in new born nursery. *Indian J Pediatr* 1983, 50: 590-594.
5. Vogell LC, Ferguson L, Gotoff SP. Citrobacter infections of the central nervous system in early infancy. *J Pediatr* 1978, 93: 86-88.
6. Graham DR, Anderson RL, Ariel FE, *et al.* Epidemic nosocomial meningitis due to *Citrobacter diversus* in neonates. *J Infect Dis* 1981, 144: 203-209.
7. Freedman RM, Ingram DL, Gross I, Ehrenkranz RA, Warshaw JB, Baltimore RS. A half century of neonatal sepsis at Yale. *J Dis Child* 1981, 135: 140-144.

## Familial Noonan Syndrome

**L. Kasturi**  
**A.V. Kulkarni**  
**V.A. Mashankar**  
**U.A. Desai**

The Noonan syndrome was first recognized as a distinct clinical entity in 1963 by Noonan and Ehmke(1). The Noonan syndrome is characterized by dysmorphic facies, congenital heart disease and short stature. It may be inherited as an autosomal dominant trait and has a wide range of severity(2). We report on four cases of Noonan syndrome

*From the Department of Pediatrics, BARC Hospital, Anushaktinagar, Bombay 400 094.*

*Reprint requests: Dr. L. Kasturi, 7-D, Everest, Anushaktinagar, Bombay 400 094.*

*Received for publication: December 7, 1993;*

*Accepted: November 4, 1994*

from three generations from one family.

### Case Reports

*Case 1:* The index patient, a baby girl was born by lower segment Cesarean section to a 27-year-old mother. Birth weight was 2.7 kg, head circumference 33 cm and length 49 cm. Antimongoloid slant, hypertelorism, epicanthic folds, bilateral ptosis, lowset ears, thick lips and single palmar crease on left hand were noticed at birth. There was no webbing of the neck or limb edema. An ejection systolic murmur was detected on 3rd day of life. There was no cyanosis and the baby was asymptomatic. The neonatal period was uneventful.

In view of the peculiar facial features and the cardiac murmur, the baby was investigated. Two dimensional echocardiography and color doppler revealed supra-avalvular pulmonic stenosis with a systolic gradient of 48 mm of Hg across the pulmonary artery. The karyotype was 46, XX. A diagnosis of Noonan syndrome was made because of