

CONGENITAL SYPHILIS — A REPORT OF 3 CASES

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SYNOPSIS

3 cases of congenital syphilis were seen in 1986 in the Paediatric Dept of Alexandra Hospital. The clinical findings and laboratory results of each case are presented and discussed. The management of the antenatal mothers and the newborn with syphilis is outlined.

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INTRODUCTION

Congenital syphilis though rare in Singapore today is a dreaded complication of syphilis. The incidence of congenital syphilis in the past years have remained low (Table 1). Although the overall incidence of syphilis has remained steady the total number of reported cases of infectious syphi-

lis is on the rise (Table 1) with the consequence that we may see more cases of congenital syphilis.

In 1986 we encountered 3 cases of congenital syphilis at the Paediatric Dept of Alexandra Hospital. In two of the cases mothers were screened antenatally for syphilis early in the pregnancy and were found to be negative. Two of the parents (1 father and 1 mother) were treated for syphilis before birth of the offsprings.

We report these cases to raise the awareness that congenital syphilis is still a problem in Singapore. Our present system of screening of antenatal mothers although comprehensive is not full proof against congenital syphilis.

Table 1
STATISTICS ON SYPHILIS IN SINGAPORE

Year	Cases of Congenital Syphilis/1000 live births	Incidence of Syphilis/100,000 population	No. of reported infectious syphilis
1980	0.15	30.9	155
1981	0.12	39.7	169
1982	0.12	39.9	285
1983	0.34	41.6	239
1984	0.29	44.0	328
1985	0.09	33.2	264
1986	0.16	35.1	391

From: The epidemiology of Syphilis in Singapore
Lee C T & Thirumorthy T
(Annual Academy of Medicine 1987)

CASE REPORTS

3 Cases of Congenital Syphilis

Case 1

Baby T.B.L. was born to 24 year old G₃P₁ mother at 31 weeks gestation at a private hospital. She was delivered vaginally and had good Apgar Score at birth. Birth weight was 1445 gm and she was transferred to Alexandra Hospital on the same day for management of prematurity.

Antenatally her mother was seen by a private obstetrician since the first trimester and was reported to be well. HBsAg and VDRL tests done in the first trimester were both negative.

Physical examination of patient on admission to Paediatric Intensive Care Unit showed that she was pink, circumference of head = 26.2 cm, BP 58/23 mmHg. Both heart sounds were heard and no murmurs were detected. There was bilateral subcostal retraction with some crepitations on auscultation. There was hepatosplenomegaly (liver 2 cm, spleen 1 cm). No petechiae or purpurs were noted.

Serial Liver Function Tests showed raised Bilirubin and Enzymes.

	TP	Alb	S Bil	SAP	SGPT	SGOT
25.11.86	5.8	3.1	8.7	243	19	52
6.1.87	4.9	3.2	7.4	531	197	237
12.1.87	5.4	3.4	7.4	549	139	175
20.1.87	4.8	3.1	4.9	407	65	102

HBsAg — negative

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A diagnosis of prematurity with hyaline membrane disease (HMD) was made. In view of the finding of hepatosplenomegaly, a diagnosis of intrauterine infection was entertained.

Investigations were carried out to confirm intrauterine infection whilst the patient received intensive care treatment for HMD.

Hemoglobin — 14.2 g/dl. Retic 2.5%

WBC 36,500/cu mm

Platelets 95,000/cu mm

Igm 426 mg/dl (N < 20)

Blood toxoplasmosis — negative

Urine for CMV — negative

Rubella titre — 32

VDRL — reactive (512)

TPHA — IgG — reactive

— IgM — reactive

Repeated mother's blood VDRL — reactive (32)

TPHA — reactive

Serial Lumbar Puncture did not show pleocytosis or elevated proteins but had a positive serology. Xrays of both knees were suggestive of intrauterine infection. She was diagnosed to have Congenital Syphilis with possible CNS involvement and given Crystalline Pen 37,500 units IM 12 hourly for 14 days.

Serial Lumbar Puncture were done:

	19.11.86	24.11.86	29.11.86	11.12.86
Appearance	—	sl. blood stained	xanthochromic	xanthochromic
Cell count	12	15	10	4
Glucose	23	66	55	69
Chloride	725	690	695	700
Total Protein	150	60	150	60
Globulins	+	trace	+	+
Others	REC-few	mainly polys few lymph	few lymph	—
Culture	NBG	NBG	NBG	NBG
VDRL) TPHA)	not done	unsuitable	reactive ⁽⁴⁾ IgM specific IgG specific	not done not reactive

* NBG — no bacterial growth

Both parents were referred to Middle Road Hospital (MRH) for treatment. The father had a history of exposure to sexually transmitted disease during his wife's pregnancy and was treated by his general practitioner. However, he was diagnosed as possibly early latent syphilis at Middle Road Hospital and given three weekly injections of Benzathine Penicillin (2-4 mega).

Baby was discharged on Day 56 of life at 2030 gms. She was followed up at the Outpatients. Her jaundice slowly resolved. Serial LFT showed improvement then became normal. Her blood VDRL and TPHA became negative. A repeat CSF examination was normal and CSF, VDRL and TPHA were negative. At her last follow up she was clinically normal and developing normally.

Case 2

Baby V was an Indian girl first seen at 2 months of age for complaints of generalised maculopapular rash of 1½ weeks duration. This was associated with peeling of skin over the palms and soles. She was brought to see a general practitioner and given some cream with no effect. On further questioning, there was a history of fever for 2 days, poor feeding, vomiting and irritability for one day. There was no history of snuffles.

Her mother had delivered a macerated stillbirth at 30 weeks previously in Alexandra Hospital. Investigations then showed VDRL positive (1:128) and FTA was borderline positive. However, her mother defaulted treatment. At her second pregnancy, baby V's mother was referred to Middle Road Hospital at 30 weeks because of positive VDRL and FTA tests. She was given 3 weeks of oral erythromycin because of a question of penicillin allergy. There were 2 episodes of antepartum haemorrhage at 32 and 33 weeks. These episodes resolved with bed rest.

Baby V was delivered at 36 weeks gestation vaginally in Kangaroo Maternity Hospital, birth weight 2.1 kg. She was well at birth and discharged after 3 days. At 6 weeks of age, baby V was seen at Middle Road Hospital. However, the blood tests done then were inconclusive and her mother defaulted further follow up. Her father was in India and hence never treated here.

Physical examination of the patient at 2 months old showed her to be febrile (T° = 37.6°) and pale. Weight was < 3 percentile (3.6 kg) and circumference of head was 35 cm (3%). There was a generalised maculopapular rash with dry scaly red maculopapular rash over the soles and palms, with extension to the forearms and shins. There was also dactylitis (swelling and warmth of interphalangeal joints) of the right middle and ring fingers. There was hepatosplenomegaly (liver 3 cm, spleen 3 cm). There was no significant lymphadenopathy and no cataracts were found. The baby was generally irritable though her muscle tone were normal and Moro's was complete. Investigations showed the following
Hb 5.9g/dl, Retics 3.2%, slight hypochromic, microcytosis
WBC 27,600/cu mm
Platelets 15,000/cu mm

Liver Function test showed hypoalbuminemia with raised SAP:-

TP — 6.0, Alb — 2.7, S Bil — 2.1, SAP — 1058

VDRL — Reactive (1:1024)

TPHA — 1st specimen — non-specific haemagglutination
2nd specimen — Reactive

Lumbar Puncture done was essentially normal —

Appearance — xanthochromic, Cells — 2, Glucose 63,

Chloride 700, Total Protein 40, globulin — negative,

culture — negative

VDRL — negative.

Xrays showed periosteal reaction in the femoral and tibial shafts and there was metaphyseal destruction about the knees, consistent with the diagnosis of congenital syphilis.

She was treated with i/m Procaine Penicillin 200,000 units for 10 days.

On follow up baby was well and development was within normal limits. Repeat VDRL was negative and TPHA was weakly reactive.

Case 3

T.M.L. was referred from MCHC for hepatosplenomegaly, noted at 3 months when she was due for her DPT vaccination. Her mother had noticed abdominal fullness since birth.

Antenatally, her mother was seen at AH and VDRL done at 20 weeks gestation was negative. Birth history was uneventful. Her mother had 2 other healthy children by her first husband with whom she had been separated for 2 years. T.M.L. is a child of her second 'marriage'.

Physical examination showed that there was bilateral cervical lymphadenopathy. There was no pallor, no jaundice and no purpura was noted. However, the abdomen was grossly distended with dilated veins with an abdominal girth of 45.6 cm. Liver was firm and palpable 9 cm below the subcostal margin with spleen palpable of 1 cm. There were desquamating erythematous rashes over the hands and feet. Investigations showed

Hb 11.4/dl, ESR 44 mm/hr

WBC 21,900/cu mm

Platelets 445,000/cu mm

Serial liver function tests showed raised enzymes (SAP, SGOT, SGPT)

Date (Normal)	S Bil (1.2)	TP (5.3-7.8)	Albumin (3.8-5.3)	AP (157-363)	ALT (9-48)	AST (16-54)
18.10.86	0.2	7.5	3.3	237	166	136
29.10.87	0.2	7.1	3.6	198	176	188

HBs Ag negative

HAV negative

Fetoprotein — negative

Rubella titre < 8

Urine CMV — positive

Blood CMV — negative

Herpes titre — 1:/64

VDRL 1/256

TPHA IgG — reactive

IgM — non-reactive

Postnatal mother's blood VDRL 1/16

TPHA reactive

HBsAg negative

Xrays of the knees showed periostitis, osteochondritis with Wimberger's sign positive (erosion of medial surface of upper tibia).

She was diagnosed to have early congenital syphilis with syphilitic hepatitis and was treated with i/m procaine penicillin 37,500 units for 10 days. On the follow up at OPD, her problem of hepatosplenomegaly was resolving. Serial liver function test showed decreasing enzyme levels and became normal at 6 months of age. A repeat blood VDRL and TPHA was negative. When last seen she was well with no splenomegaly and was developing normally.

DISCUSSION

Congenital syphilis is contracted from the infected mother through the transplacental route. The risk of transmission is highest in the first 2 years of maternal syphilis. The organisms reach the foetus via the placenta and umbilical vein. The liver is the primary site of infection with secondary spread to the skin, mucous membrane, bones and central nervous system.⁽¹⁾

The three cases reported serve to illustrate that congenital syphilis is still a problem as the overall management of syphilis in the community is not comprehensive.

The father of Case 1 was treated by a medical practitioner after he was exposed and was told he was cured. The fact that he did transmit syphilis to his wife resulting in his daughter having congenital syphilis and that he now has early latent syphilis indicated that he was inadequately treated.

Mothers of Case 1 and 3 were both screened antenatally for syphilis in early pregnancy. Both were negative. However, both were positive later indicating that they contacted the disease in the later part of pregnancy. This illustrates that a single VDRL in early pregnancy (screening method used in all Government Hospitals and by most private Obstetricians) alone, would not detect all cases of syphilis in pregnancy. It is suggested that a repeat be done in later part of pregnancy whenever a medical or epidemiological risk factor is recognised.

Erythromycin treats antenatal mothers adequately but there have been reports that it fails to treat foetal infection adequately as Erythromycin's passage through the placenta is unpredictable. Case 2 illustrated this problem very well. During mother's second pregnancy at 30 weeks gestation she was given 3 weeks of Erythromycin because of a suspicion of Penicillin allergy. This did not prevent Case 2 from contracting syphilis. She had an inconclusive serological titre at 6 weeks of age but the fact that mother was treated with Erythromycin warranted her to have a course of treatment for congenital syphilis but this was not given. Thus all babies born to antenatal mothers with syphilis treated with any drug other than Penicillin especially Erythromycin should be worked up and treated with Penicillin after delivery.^(2,3)

The drug of choice for treatment of syphilis is Penicillin. **Specific therapy for those with primary, secondary and early latent syphilis of less than 1 year duration (including antenatal mothers) is as follows:**

- a) **Benzathine Penicillin G** 2.4 mega units IM (1.2 mega in each buttock).
A repeat dose can be given a week later. (A total dose of 4.8 mega).
- b) **Acqueous Procain Penicillin G** 600,000 units/day IM for 10 days.
(A total dose of 6 mega units).
- c) **If Penicillin allergy is present**
 - i) Erythromycin (oral) 500 mg 6 hourly for 15 days or
 - ii) Tetracycline hydrochloride (oral) 500 mg 6 hourly for 15 days or
 - iii) Doxycycline (oral) 100 mg twice a day for 15 days.

For those with syphilis of more than 1 year duration including antenatal mothers, Penicillin is again the drug of choice.

- a) **Benzathine Penicillin** 2.4 mega IM weekly for 3 successive weeks. (A total of 7.2 mega units)

- b) Aqueous Procain Penicillin G 6 00,000 units IM daily for 21 days. (A total of 12.6 mega units).

If Penicillin allergy is present

- a) Erythromycin (oral) 500 mg 6 hourly for 30 days or
 b) Tetracycline (oral) 500 mg 6 hourly for 30 days
 (NB: Tetracycline if given to antenatal mothers can cause staining of teeth of the baby and is thus not recommended).

A VDRL (non-treponemal test) is done on cord blood for screening of newborns for congenital syphilis. A negative result practically rules out congenital syphilis. If a positive result is obtained a treponemal specific test, Fluorescent treponemal antibody abs (FTA-ABS) or Treponema pallidum hemagglutinating antibody (TPHA) should be done. Both tests are extremely sensitive and very specific. The indirect immunofluorescent test FTA-I gM for detection of specific IgM antibodies to T. pallidum has a high false negative rate (20-40%) in infants with congenital syphilis whose onset of disease is late and this alone should not be used for the diagnosis of congenital syphilis.⁽⁴⁾

The decision to treat a neonate for congenital syphilis is based on clinical presentation, maternal history of previous positive serology and antisyphilitic treatment and on results of serological testing of newborn and mother.

Babies with positive serology or have signs and symptoms of congenital syphilis are to be treated. A mandatory lumbar puncture is performed as treatment depends on whether there is evidence of Central Nervous System infection (CSF pleocytosis, elevated protein and positive serology). The following regime is adopted in the Paediatric Dept, Alexandra Hospital.

If there is no CNS infection

- a) Procaine Penicillin G 50,000 units/kg IM in a single daily dose for 10 days, OR
 b) Benzathine Penicillin G 50,000 units/kg IM in a single dose has been recommended as adequate therapy but lack of penetrating of the CSF by this dose and the difficulty in interpretation of CSF in the newborn, it is not recommended^(5,6) except in cases where the patient is unreliable and may not return for further therapy as in our Case 2.

Those with CNS infection should be treated with

- a) Crystalline Penicillin G 50,000 units/kg/day IM in 2 or 3 divided doses for 2 — 3 weeks. OR
 b) Procain Penicillin 50,000 units/kg in one daily dose for 2 — 3 weeks.^(5,7,8)

Babies with positive serology that need not be treated are

- 1) those clinically well, ie no signs of congenital syphilis or intrauterine infection whose mothers were adequately treated (document treatment) with Penicillin.

- 2) Well babies with lower VDRL titers than that of mothers who have been adequately treated in the past. In these situations the positive serology indicates transplacental passage of maternal antibodies. These babies should be followed up, however, and a repeat serology at 6 months by which time they should be negative as mother's antibodies should have disappeared. When in doubt it is best to consult the paediatrician or dermatologist.

Often an infant will be treated despite uncertain evidence of syphilis, before confirmatory evidence can be obtained, in an effort to prevent further progression of congenital syphilis.

During the acute stage, nasal secretions and open syphilitic lesions are very infectious. Precautions should be taken by health care personnel handling these babies. Twenty-four hours after starting Penicillin therapy the infant is no longer infectious. These babies should also be screened for evidence of other sexually transmitted disease like Gonorrhoea, Chlamydia and Hepatitis B.

All babies should be followed up with serologic: I test at 2, 4, 6 and 12 months. Any rise in titre is an indication for treatment. The non-treponemal tests (eg VDRL) will become negative in 90% of those adequately treated. The FTA-Abs test may remain positive. Those with neurosyphilis should have serological and spinal fluid examination done monthly for three months and then every 6 monthly for 3 years. If the infant is adequately treated the cell count and protein will become normal in 2-3 months and the non-treponemal titres will fall. The FTA-Abs may remain positive. Any rise in titres or change in cell count is an indication for retreatment.

Congenital syphilis is a notifiable infectious disease. Adequate notification will allow epidemiological measures to be instituted. The parents of the baby with congenital syphilis must be examined and investigated and treated when necessary for sexually transmitted diseases.

Congenital syphilis is best treated early and adequately for minimal sequelae. It can have disastrous effects if the infant is left untreated. The best and most effective method of control of this disease is prevention by education, early and adequate treatment of the adults who are exposed and effective screening and treatment of antenatal mothers.

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