Staphylococcus rose steadily after the preschool years. None were found in the first 4 years of life, but the incidences for the second and third quinquennium were 4·9% and 16·8% respectively.

Cystic fibrosis series. The series of 48 cases of CF (38 surviving) treated with continuous cloxacillin for from 1 to 11 years since 1964, showed in January 1975 (or shortly before death before this date) the differences from the normal series which are shown in the Table. There were no significant differences in respect of Pneumococcus, Proteus, and Klebsiella, and the CF series were not tested against the haemolytic streptococcus.

Discussion
In the normal population no precipitins were found in the first year of life. No staphylococcal precipitins were found in the first 4 years, but subsequently the incidence rose from 4·9% in the second quinquennium to 16·8% in the third quinquennium. Precipitins against the pneumococcus were 2% in the second and 5·3% in the third quinquennium. The incidence of haemophilus precipitins was between 2·1% and 3·4% in all quinquennia. Precipitins against other organisms are rare, with the exception of haemolytic streptococcus which showed an incidence of 3·4% in the second quinquennium only.

The CF figures are markedly different. The well known predisposition of CF to H. influenzae and Ps. aeruginosa infection is confirmed, but it should be noted that cases in the third quinquennium have not had the benefit of continuous antistaphylococcal therapy since early diagnosis. Cases of CF are well known to be particularly prone to lung damage from staphylococcal infection. The present findings indicate that the development of staphylococcal precipitins can be prevented by continuous cloxacillin therapy, and that pre-existing staphylococcal precipitins disappear slowly on this treatment.

Summary
Well children show a steady increase of staphylococcal precipitins throughout life. Lower incidences are found for precipitins against H. influenzae, Pneumococcus, and haemolytic streptococcus. Precipitins against Proteus, Pseudomonas, and Klebsiella are rare. There are marked differences between the normal children and the cloxacillin-treated children with cystic fibrosis in respect of Pseudomonas and H. influenzae which rise to over 50% of cystic fibrosis children in the third quinquennium. However, the development of staphylococcal precipitins can be prevented by continuous antistaphylococcal therapy.

We acknowledge the help of the late Dr. R. L. Newman, and thank the Cystic Fibrosis Research Trust for the grant which financed this investigation.

Frequency of other malformations in congenital hypoplasia of depressor anguli oris muscle syndrome
The interest in the asymmetric crying facies due to congenital hypoplasia of the depressor anguli oris muscle (DAOM) has grown in recent years because of its association with congenital abnormalities and, more specifically, with congenital heart disease (Cayler, Blumenfeld, and Anderson, 1917). Papadatos et al. (1974) have reported on the possibility that the DAOM hypoplasia is a genetically determined condition. This paper compares the frequency of congenital abnormalities in neonates with and without DAOM hypoplasia.

Material and methods
All newborn infants born between 1 August 1972 and 1 December 1973 at Alexandra Maternity Hospital were specifically examined for the presence of DAOM hypoplasia or for other congenital abnormalities. There were 6487 neonates, 3314 males and 3173 females. DAOM hypoplasia was observed in 44 newborns; on all of whom x-rays of the heart and of the whole skeleton were taken, as well as an electrocardiogram. One year after delivery 32 of the 44 neonates with DAOM hypoplasia were located and examined for the purpose of detecting additional congenital anomalies. A complete physical and cardiac examination was done, as well as a careful urinalysis and a BM nitrit test (Fuchs and Gutensohn, 1967) for detection of possible pyelonephritis due to anomalies of the urinary tract. In the group of newborns without DAOM hypoplasia 29 suffered from congenital heart disease. 19 of them were
re-examined after a year. Of the remaining 10, 3 could not be located and 7 had died due to their congenital heart disease.

Results were analysed by the $\chi^2$ test with Yates's correction.

**Results**

**Affected neonates.** Of the 6487 newborn infants examined 44 were found to have DAOM hypoplasia, confirmed in 15 of them by electromyographic studies. The mean gestation of the affected newborns was 39.6 weeks (range 32–42 weeks) and the mean birthweight 3283 g (range 1350–4259 g). In no case was there consanguinity of the parents. The left side was affected in 30 cases and the right in 14. The ensuing asymmetry was characterized in 10 neonates as severe, in 17 as moderate, and in 17 as mild. 24 of the affected neonates were girls and 20 were boys.

Congenital anomalies were detected in 9 of the 44 newborns with DAOM hypoplasia, an incidence of 20%. Of these, 3 had congenital cardiac abnormalities (septal defects), 2 flat hemangiomata, one undescended testis, one micrognathia and asymmetry of the maxillae, one congenital dislocation of the hip, and one supernumerary breast nipple on the left side.

Examination of the 32 infants at the age of one year showed 2 additional cases with congenital dislocation of the hip.

**Unaffected neonates.** Of the 6443 unaffected neonates, 176 were found to have congenital abnormalities (2.7%) (Table). 9 of them had

### TABLE

**Congenital abnormalities in the unaffected neonates**

<table>
<thead>
<tr>
<th>System</th>
<th>No. of cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Circulatory</td>
<td>29</td>
<td>0.45</td>
</tr>
<tr>
<td>Urogenital</td>
<td>57</td>
<td>0.88</td>
</tr>
<tr>
<td>Bone-muscles</td>
<td>43</td>
<td>0.67</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>9</td>
<td>0.14</td>
</tr>
<tr>
<td>CNS</td>
<td>7</td>
<td>0.11</td>
</tr>
<tr>
<td>Respiratory</td>
<td>1</td>
<td>0.02</td>
</tr>
<tr>
<td>Skin</td>
<td>9</td>
<td>0.14</td>
</tr>
<tr>
<td>Multiple congenital abnormalities</td>
<td>9</td>
<td>0.14</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>12</td>
<td>0.19</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>176</strong></td>
<td><strong>2.73</strong></td>
</tr>
</tbody>
</table>

more than one abnormality. Congenital heart anomalies were diagnosed in 29 neonates: ventricular septal defect 14, ductus arteriosus 2, Fallot's tetralogy 2, and one each of stenosis of the pulmonary artery, coarctation of the aorta, aortic stenosis, atrial septal defect, Ebstein syndrome, and common arterial duct with hypoplasia of left heart. In 5 cases the exact nature of the congenital heart anomaly was not diagnosed.

**Statistical analysis.** The observed difference in the frequency of congenital abnormalities between the infants affected with DAOM hypoplasia (20%) and the unaffected ones (2.7%) is very significant ($\chi^2=43.7$, $P<0.001$). 3 cases of congenital cardiac anomalies were observed in the first group (6.8% frequency), while 29 such cases (0.45%) were diagnosed in the second group. The observed difference in frequency was again significant ($\chi^2=24.29$, $P<0.001$).

**Discussion**

The incidence of congenital malformations in an unselected population varies from 1-24% to 5% (Stevenson, Worcester, and Rice, 1960), depending on the criteria used (Craig, 1969; Pantelakis, Karageorga-Labama, and Bartsockas, 1973). When only the serious congenital abnormalities are included, the incidence varies between 0.67 and 2.1% (Malpas, 1937; McIntosh et al., 1954; Marden, Smith, and McDonald, 1964).

Cayler et al. (1971) and Pape and Pickering (1972) reported a high incidence of congenital abnormalities in infants with DAOM hypoplasia (59% and 86% respectively). But their material was a selected one, as it came from hospitalized or institutionalized infants and children. Our own research was done on a routine neonatal population and our findings of 20% of the infants with DAOM hypoplasia having congenital malformations seems to be reliable. 2.7% of the control neonatal population had congenital malformations. The low incidence of associated congenital anomalies (only 2) found by Perlman and Reisner (1973) among the 41 newborns with asymmetrical crying facies studied may be due to the fact that their babies were not thoroughly investigated (Gellis, 1975). The frequency of congenital cardiac malformations in a normal neonatal population has been found to be 0.1-0.9% (Craig, 1969). In our own patients the frequency was 0.45%, while the observed frequency of congenital cardiac malformations among newborns with DAOM hypoplasia was as high as 6.8%.

Thus it can be concluded that an eightfold increase of associated anomalies was observed in the presence of DAOM hypoplasia. The practical implication from the present study is that newborns with DAOM hypoplasia should be examined for other congenital abnormalities, particularly of the cardiovascular system.
Summary

6487 newborns, the products of consecutive deliveries, were specifically examined in the present study for congenital abnormalities. 44 of them presented hypoplasia of the depressor anguli oris muscle (DAOM). Associated congenital malformations were compared to those observed in the general neonatal population. The frequency of congenital anomalies among infants with hypoplasia of DAOM was 20%, as compared to 2-7% in the control group of the same period. These figures indicate an eightfold increase of associated anomalies in the presence of the hypoplastic DAOM. Cardiovascular anomalies were found in 6-8% of the affected newborns, as compared to 0-45% in the control group. Newborns with hypoplasia of the DAOM should be examined for other congenital anomalies, particularly of the cardiovascular system, since these tend to occur at a significantly higher frequency.

References


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Value of cystography in urinary tract infections

It is customary in paediatric practice to investigate the urinary tract radiologically by means of an intravenous urogram (IVU), and a micturating cystogram when a child is first diagnosed as having a urinary tract infection (UTI) whether symptomatic (Smellie, 1969; Rolleston, Shannon, and Utley, 1970) or asymptomatic (Davies et al., 1974; McLachlan et al., 1975). However, cystography may be associated with numerous complications. 13 are listed by McAlister, Cacciarelli, and Shackelford (1974), and after one of our patients required a general anaesthetic for removal of a catheter, we decided to see whether in patients with a normal IVU, cystography added any useful information.

Patients and methods

X-rays of all the children with a bacteriologically proven UTI, who had had both an IVU and a micturating cystogram over a 2-year period, were reviewed. The diagnosis of UTI was made if a fresh specimen of urine contained a single organism with a quantitative count of 100 000 organisms/ml or more. For the IVU, after a plain film of the whole abdomen had been taken, an injection of Urovison (sodium and methylglucamine salts) 1.0 ml/kg body weight was given. No abdominal compression was used.

The first film was taken at 5 minutes (3 minutes in very young children). If the 5-minute film showed two kidneys filling well, a bottle of cold Pepsi-Cola was given through a straw with the child lying down. This distended the stomach and showed the kidneys through a gas filled stomach. At 15 minutes a complete renal tract film was taken. This was occasionally supplemented with an oblique film to show, if necessary, a kidney partially obliterated by overlying bowel shadow, and to bring it into the area of the clear gas-filled stomach.

If an adequate film of the bladder was obtained, a further renal tract film was taken after micturition to assess bladder emptying. Tomography was not used in any of these patients.

Cystourethrography was performed on another day. The patient was catheterized with a Foley's catheter (a self-retaining catheter was used as it had been found that some children would extrude a fine polyethylene catheter before adequate bladder filling was obtained). A specimen of urine was sent for culture, but the bladder was not emptied. The subsequent examination was controlled by minimal intermittent screening. The bladder was filled by a 60 ml catheter-tip syringe with half diluted Urovison and sterile water, or if the bladder was very full with an undiluted Urovison. If reflux was seen to occur no further contrast was added, otherwise the bladder was filled to tolerance.

The amount of contrast introduced varied between 25 and 75 ml according to bladder volume and the age of the child. No screening films were taken unless reflux was
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Arch Dis Child 1976 51: 891-893
doi: 10.1136/adc.51.11.891

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