

# OMPHALOCELE

## A Review of 10 Cases

by

A. M. VARE, M.B.B.S., M.Sc. (Medical)

and

P. C. BANSAL, M.S., F.R.C.S.

Omphalocele is one of the serious malformations of a newborn baby. It is associated with high mortality which may be further increased in large percentage of those infants with coexistent anomalies and who are born prematurely. There is herniation of abdominal viscera in the umbilical cord. The sac consists of two layers of tissues, the inner peritoneum and the outer, amnion which fuse together to form a thin membrane.

Smith *et al* (1966) have classified omphalocele as small and large. Small omphalocele or hernia into the umbilical cord contain intestines but never liver or other organs. The umbilical cord is usually attached at the apex of the sac. Large omphalocele usually contains liver, small and large intestines.

*Embryology:* In the third week of intrauterine life, four embryonic folds (a head fold, a tail fold and two lateral folds) grow and meet in the middle line anteriorly, forming the future umbilical ring. At the umbilical ring, the somatic layers of these folds are continuous with the amniotic sac which covers the outside of the umbilical cord. Incomplete growth of these folds produce a defect at

the future umbilicus. This defect is bridged by the amniotic sac with which the folds are continuous. The intestines and other viscera, therefore, herniate into the amniotic sac and can be visualized through the transparent membrane.

### *Observations*

#### *Age and Sex*

It was observed that large number of omphaloceles were associated with prematurity. There were 2 full term babies and 8 were premature. Of 10 omphalocele babies, 5 were males and 5 were females (Table-1).

#### *Size of defect*

This was a measurement of a gap in the abdominal wall, the diameter of the sac itself was much greater. The facial defect (gap-diameter) is smaller and varied from smallest 1 cm. to greatest 10 cm. The diameter of 3 cm. was maximum seen in small omphalocele, whereas all those over 3 cm. were large omphaloceles. In recent state the omphalocele sac is thin and translucent so that the sac contents can be identified by external examination, but after a few hours the sac becomes thicker, lusterless and opaque. It is evident that the smaller the sac the better are the chances of survival.

\*From Dept. of Anatomy, Medical College, Aurangabad.

Received for publication on 24-10-1970.

TABLE I

Foetus No.	Age in weeks	Sex	Site of defect	Size of defect	Contents of sac	Sac intact or ruptured	Attachment of cord.
1	34	M	Upper abdomen	Sac 7×6 cm.	Liver, small and Large intestine	Intact	Upper & left part of sac
2	36	M	Middle abdomen	4×4cm.	Liver, small intestine	Intact	Lt & lower part of sac
3	36	F	Middle abdomen	3 × 3 cm.	Small & large intestine, malformation of gut	Intact	Do.
4	28	M	Almost whole of ant. abd. wall absent	8 × 7 cm.	Liver, spleen small and large intestine & malrotation	ruptured	above pubic symphysis
5	29	F	Do.	7 × 7 cm.	Liver, small & larger intestine & urinary bladder	ruptured	at the margin to the left side
6	28	M	Upper & right abdominal wall absent	10 × 6 cm.	Liver, intestine stomach, spleen, appendix	Intact	above pubic symphysis
7	28	F	Umbilical region	2 × 2	Small intestine	Intact	at the apex of sac
8	32	F	Ant. Abd. wall absent	8 × 8cm.	Liver, small & large intestines malformation	Intact	at pubic symphysis
9	32	M	Ant. Abd. wall absent	8 × 10 Cm.	Liver, intestine stomach, ectopia viscae, agenesis of hind gut	Intact	near xyphoid process
10	28	F	Umbilical region	1 × 1	Intestine	Intact	at the apex of sac.

Abbreviations : M—Male, F—Female, ant. abd. wall—anterior abdominal wall.

*Content of sac (Table 1)*

By far the commonest content of the sac was midgut loop (Fig. 2) or midgut loop with liver (Fig. 3). At times other viscera of abdomen namely large intestine, stomach, spleen and urinary bladder were found in the hernial sac along with the midgut loop and liver. It is interesting to note that the omphalocele of supraumbilical region usually contain liver, small intestines and stomach. Omphalocele of infraumbilical region contained urinary bladder and large intestine (Fig. 4).

*Sac rupture*

It is evident that rupture of sac is a grave complication of omphalocele because of secondary changes which occur in the eviscerated bowels and infection. Of 10 sacs, 2 were ruptured and the remaining 8 were unruptured. Ruptured sacs were found in large omphaloceles.

*Attachment of cords*

Umbilical cord is either attached to the apex of the sac, or at any point at the margin of the sac. It was attached to the apex of sac in 2 cases (Fig. 1), at the mar-

gin of sac near pubic symphysis in 2 cases (Fig. 4), at the upper and left margin in 4 cases and at the lower and right margin in the remaining 2 cases. Attachment of umbilical cord to the apex of the sac was observed in small omphalocele and as a rule at the margin of the sac in large omphalocele.

#### Coexistent Malformations (Table II)

Of the 10 cases, there were 2 in whom omphalocele was the only malformation. Each of the other 8 cases had one or more coexistent malformations. Serious congenital malformations like spina bifida, anencephaly (Fig. 1), scoliosis of vertebral column and anomalies of the limbs were found in large number of cases (Figs. 4, 5). Less serious defects like hare-lip, meningocele, diaphragmatic hernia and genito-urinary anomalies were noticed in a few cases (Fig 4). Malrotation and malformation of gut was observed in 6 cases, extrophy of bladder in 2 cases and agenesis of hindgut in 1 case (Table 1).

TABLE II  
Showing coexistent malformations

Malformations	No. of cases
Spina bifida	4
Anencephaly	4
Scoliosis of vertebral column	3
Anomalies of limbs	7
Hare-lip	1
Meningocele.	2
Urinary anomalies	2
Diaphragmatic hernia	2
Anomalies of uterus	2

TABLE III  
Showing maternal age, parity and complications during pregnancy

Case No.	1	2	3	4	5	6	7	8	9	10
Age of mothers in years	25	23	20	25	18	28	20	20	30	18
Parity of mother	Primi	2nd	2nd	primi	7th	2nd	primi	6th	primi	primi
Other complications	Hydr- amnios A.P.H.	Hyder- amnios	Breech	Hyd- eram- nios	A.P.H.	Trans- verse- lic	Trans- verse- lic	...	...	...

Abbreviations: A.P.H.—Antepartum haemorrhage, Trans. lie—Transverse lie.

All cases of scoliosis of vertebral column were associated with large omphalocele probably because the defect in the abdominal wall is stretched out.

#### Mothers of omphalocele babies

##### (a) Age and parity

Mothers of these ten cases were between the ages of 18 years to 30 years, the average being 22 years. Of 10 mothers of omphalocele babies, 5 were primiparas, 3 were second paras, and remaining 2 were 6th and 7th para. It appears therefore that there is some relationship between age and parity of mother to omphalocele (Table III).

##### (b) Other complications during pregnancy

Of the ten mothers of omphalocele babies, 3 had no complication during pregnancy, but the rest, 7 mothers, had either hydramnios, antepartum haemorrhage or abnormal presentation of the fetuses (Table III).

#### Discussion

##### Incidence

Mckeown (1953) has reported 1 omphalocele in 3200 births, whereas O'leary and Clymer (1941) found incidence of omphalocele, 1 in 3,300 births, Jarcho (1937) 1 in 6,600 births and Hutchin and Goldenberg (1965) 1 in 5000 births. In the present series the incidence of omphalocele was found to be much higher (1 per 1500 births).

Prematurity and Sex

TABLE IV  
Showing the incidence of prematurity and sex

	Izant et al (42 cases).	Hutchin & Golden berrg (13 cases).	Moore (29 cases).	Present series 10 cases.
Prematurity	16	6	....	8
Sex	Male 27 Female 15	—	Male 19 Female 10	Male 5 Female 5

Izant *et al* (1966) and Hutchin and Goldenberg (1965) have found no significant difference in incidence of omphalocele and prematurity.

The findings of the present work regarding prematurity and incidence of omphalocele are not in agreement with those of other workers.

Izant *et al* (1966) and Moore (1963) found that omphalocele occurred much more frequently in males than in females. In the present study no such relationship was observed.

Size of defect and sac

The size of abdominal defect varied. It had ranged from a small defect to from xyphoid to pubis defect which was reported by Brezin and Mayer (1953). Jones (1963) holds the view that there is some correlation between the large defect, herniation and prematurity. Soper and Green (1961) reported that the size of sac varied from 2 cm. to 14 centimeters. In the present series the defect varied from

1 cm. to 10 cms. (Table 1). In 3 cases the defect was so large that almost whole of anterior abdominal wall was involved.

Majority of defects occurred to the right of umbilical cord (Moore, 1963). In his series of 31, in 25 cases the defect was to the right of the cord and in 6 it was to the left. In the present series the defect was found to the right of the cord in 4 cases and to the left in 2 cases.

Associated Malformations

Moore (1963) reported gross associated congenital malformations in 3 out of 31 omphalocele babies. Jones (1963) found majority of omphalocele babies (35 out of 45 babies) coexistent major malformations. Moore (1963) and Jones (1963) observed absence of intestinal malformation and fixation in large number of omphalocele babies. Diller and Travis (1955) reported a portion of urinary bladder eviscerated in omphalocele.

It is evident from table-V that the findings in the present series more or less agree with those of the other workers.

TABLE V  
Showing the comparative data of various workers of coexistent malformations

	Extrophy of Bladder	Malro- tation	Agene- sis of hindgut	Skeletal	Diaphrag- matic hernia.
Hutchin and Goldenberb (13 cases)	2	7	1	1	2
Izant et al (42 cases)	4	3	3	3	1
Present series (10 cases)	2	3	1	2	2

**Summary**

A total of 10 foetuses with omphalocele were met with over a period of 7 years.

The incidence of omphalocele was 1 per 1500 births which is significantly higher than that reported by other workers.

Maternal age and parity bear a definite relation with the incidence of omphalocele. High incidence was found in mothers of younger age group and in mothers of first and second para.

Omphalocele babies occurred much more frequently in premature than in full-term babies. The preponderance of male over female was not found.

Associated anomalies were present in 80% of cases. Anomalies were most frequent in gastrointestinal, genitourinary and skeletal system.

**Acknowledgement**

We are greatly thankful to Dr. M. R. Dhamdhare, Dean, Medical College, Au-

rangabad, for his permission to publish this paper.

**References**

1. Brezin, D. and Mayer, R. A.: *Surgery*. 33: 901, 1953.
2. Diller, W. E. and Travis, B. W.: *Ohio State Med. J.* 51: 756, 1955.
3. Hutchin, P. and Goldenberg, I. S.: *Arch. Surg.* 90: 22, 1965.
4. Izant, R. J., Brown, F. and Rothmann, B. F.: *Arch. Surg.* 93: 49, 1966.
5. Jarcho, J.: *Surg., Gynec. & Obstet.* 65: 593, 1937.
6. Jones, P. G.: *Arch. Dis. Child.* 38: 180, 1963.
7. Mckeown, T. J. Mac Mahon, B. and Record, R. G.: *Am. J. Hum. Gen.* 5: 168, 1953.
8. Moore, T. C.: *Ann. Surg.* 158: 263, 1963.
9. O'leary, C. M. and Clymer, C. E.: *Am. J. Surg.* 52: 38, 1941.
10. Smith, W. R. and Leix, F.: *Am. J. Surg.* 111: 450, 1966.
11. Soper, R. T. and Green, E. W.: *Surg. Gynec. & Obstet.* 113: 501, 1961.

*See Figs. on Art Paper VII-VIII*