

## Risk Factors Associated with Postoperative Outcome of Congenital Anomalies

Harsali F. Lampus,<sup>1,2</sup> Candy,<sup>1,2</sup> Leo Rendy,<sup>1,2</sup> Sabrandi P. Saputra<sup>1</sup>

<sup>1</sup>Department of Surgery, Faculty of Medicine, Universitas Sam Ratulangi, Manado, Indonesia

<sup>2</sup>Division of Pediatric Surgery, Department of Surgery, Universitas Sam Ratulangi, Manado, Indonesia.

Email: [harsali\\_lampus@unsrat.ac.id](mailto:harsali_lampus@unsrat.ac.id)

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**Abstract:** Mortality rate of newborns with congenital abnormalities worldwide is still high in the first four weeks after birth every year. This study aimed to obtain the profile of patients with congenital anomalies related with mortality after surgery at the Pediatric Surgery Department, Prof. Dr. R. D. Kandou Hospital, Manado, Indonesia. This was a descriptive and retrospective study. There were 98 samples during 2019 to 2021. Dependent variable was postoperative mortality outcome and independent variable risk factors were sex, gestational age, birth weight, and length of stay. Data were analyzed using univariate method. The results showed that postoperative death was 27.6%, and sex ratio was equally distributed (male 51%; female 49%). Gestational age was predominantly term 77.5% vs preterm 22.5%. Birth weight of  $\geq 2500$  g was 77.5% and low birth weight ( $< 2500$  g) was 22.5%. Hospitalization length of stay probably less than two weeks was 82.7%. Pediatric surgical congenital anomalies were as follows: Hirschsprung Disease (15.1%), anorectal malformation (41.7%), lateral inguinal hernia (11.3%), diaphragm hernia (3.1%), omphalocele (2.1%), gastroschisis (2.1%), undescended testis/UDT (4.1%), invagination (4.1%), duodenal atresia (3.1%), ileojejunal atresia (4.1%), esophageal atresia (5.1%), biliary atresia (1%), and mechanical bowel obstruction (3.1%). In conclusion, in the last two years, pediatric surgery services in surgical congenital anomalies had overall mortality rate of 27.6% without sex predominantly. Most patients were term babies with normal birth weight and relatively short length of stay.

**Keywords:** congenital anomalies; postoperative outcome

### INTRODUCTION

Congenital anomalies are abnormalities of babies from birth with defects or impaired function in certain organs or parts of the body. These abnormalities begin prenatally due to defects in embryogenesis or intrinsic abnormalities during fetal development. Congenital abnormalities are related to genetic factor, and also external environment. Many babies are born with serious defects due to post-conceptional disorders caused by exposure to environmental agents that are teratogenic, such as alcohol consumption, rubella, syphilis, and iodine deficiency which may interfere with fetal development.<sup>1-5</sup>

Mortality rate for newborns with congenital abnormalities worldwide is around 303,000 babies in the first 4 weeks after birth

every year. Basic Health Research report in Indonesia (*Riset Kesehatan Dasar*) 2016 stated that 1.4% of newborns aged 0-6 days of birth and 19% of newborns aged 7-28 days died due to congenital abnormalities.<sup>6</sup> Data of the World Health Organization South-East Asia Region (WHO SEARO) in 2010 estimated that the prevalence of congenital abnormalities in Indonesia was 59.3 per 1000 live births. If every year 5 million babies are born in Indonesia, then there will be around 295,000 cases of congenital abnormalities per year.<sup>7</sup>

If a baby suffers from a congenital abnormality that interferes with organ function, this abnormality will greatly affect the baby survival, especially if the abnormality requires surgery. It is understandable that the risks will

be suffered by the baby. Therefore, if surgery is needed, it should be distinguished whether the surgery must be immediate to save the baby's life (cito), or the surgery can still wait for optimal conditions (urgent), or even the surgery can be planned and wait until the condition is good (elective).<sup>8</sup> The purpose of this study is to obtain the profile of patients with congenital anomalies related to mortality after surgery at the Pediatric Surgery Department, Prof. Dr. R. D. Kandou Hospital, Manado, Indonesia.

**METHODS**

This was a descriptive and retrospective study. Samples of postoperative outcome of congenital anomaly cases were based on medical records from 2019 to 2021 at Prof. Dr. R. D. Kandou Hospital, Manado, totaling 98 cases. Dependent variable was postoperative outcome and independent variables were patients' profiles such as sex, gestational age,

birth weight, and length of stay. Data were analyzed using univariate methods.

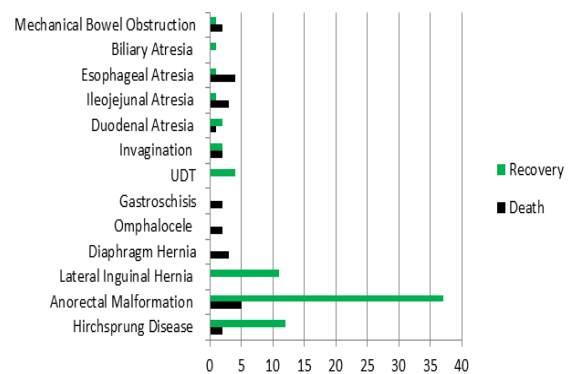
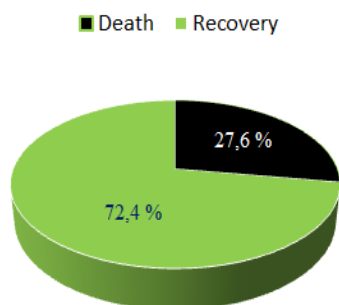
**RESULTS**

Figure 1 showed that there were 72 recovery cases (72.4%) and 26 death cases (27.6%) of congenital anomaly post operative outcome.

Figure 2 showed that sex presentation was equally distributed which was 50 male cases (51%) and 48 female cases (49%). However, cases of Hirschsprung Disease, undescended testicle (UDT), and ileojejunal atresia were most common in males in the period of 2019 to 2021 at Prof. Dr. R. D. Kandou Hospital Manado.

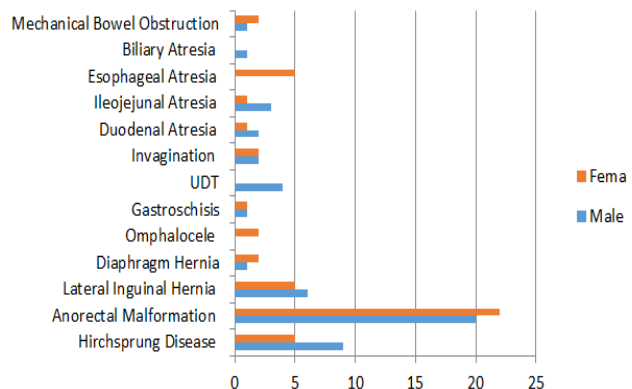
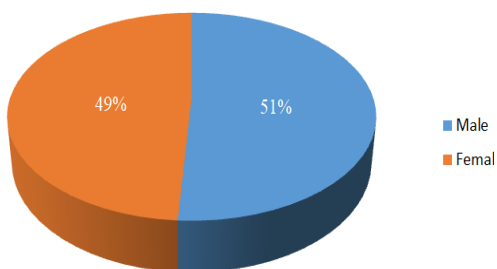
Figure 3 showed that the presentation of gestational age was 76 cases (77.5%) of term babies and 22 cases (22.5%) of preterm babies. All cases of gastroschisis were preterm babies.

**Post Operative Outcome**



**Figure 1.** Postoperative mortality and distribution of congenital anomaly cases

**Sex**



**Figure 2.** Presentation of sex distribution of congenital anomaly cases

Figure 4 showed the distribution of patients' birth weights. Patients with  $\geq 2500$  gram birth weight were 76 cases (77.5%) and patients with  $< 2500$  gram birth weight were 22 cases (22.5%). Based on the data, all cases of gastroschisis had birth weight less than

2500 grams.

Figure 5 showed that hospitalization length of stay for  $\geq 14$  days was 17,3% of cases (16 patients) and for  $< 14$  days was 82,7% of cases (82 patients).

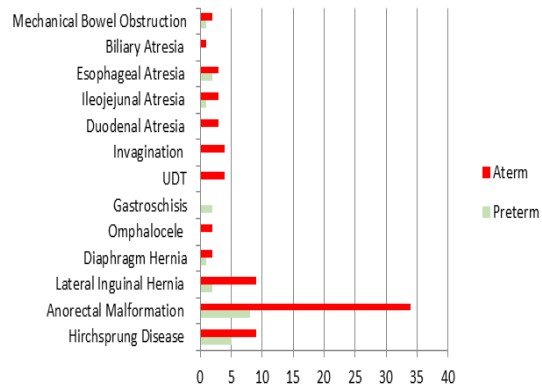
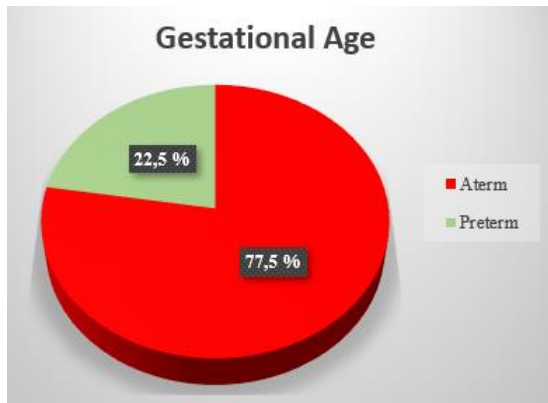


Figure 3. Presentation of gestational age and distribution of congenital anomaly cases

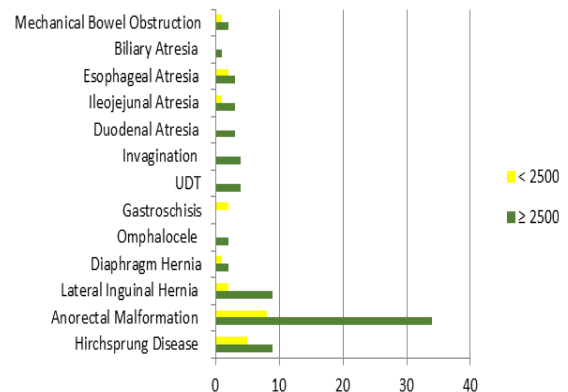
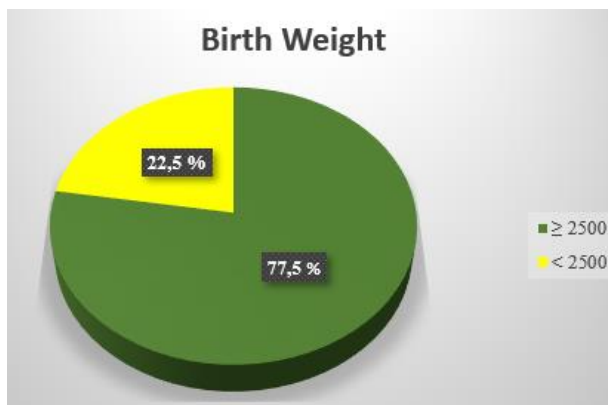


Figure 4. Presentation of birth weight and distribution of congenital anomaly cases

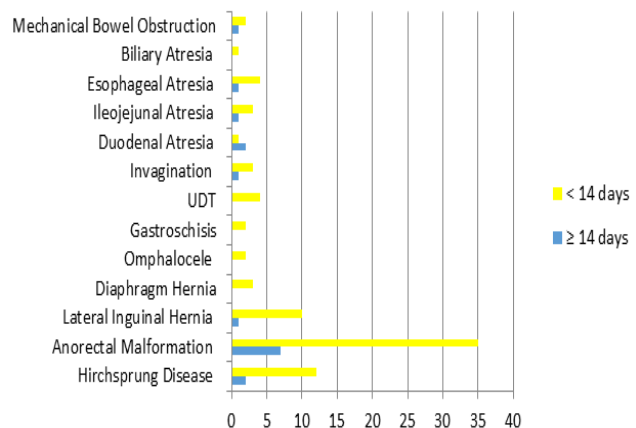
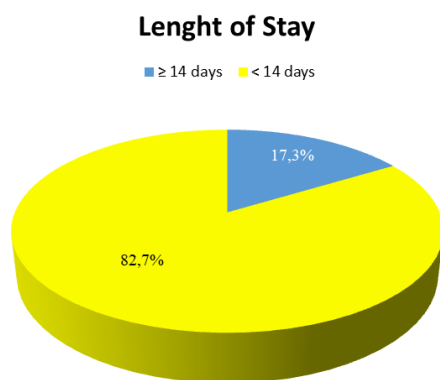


Figure 5. Presentation of length of stay and distribution of congenital anomaly cases

## DISCUSSION

Etiology factors of congenital anomalies were unknown in 40%-60% cases. However, it is thought that gene mutation is the most influential factor in the occurrence of congenital anomalies. Among the several etiologies of genetic disorders, chromosomal abnormalities reach 6%, abnormalities single gene 25%, and multifactorial 20-30%. Marriage between family is considered as one of the important factors that contributes to the incidence of congenital anomalies. There are several risk factors that support the occurrence of congenital anomalies, as follows: genetic disorder, environment, infection, nutritional status, socioeconomic status, and demographics.<sup>3</sup>

It is estimated that 3% of newborns could have congenital anomalies. Congenital gastrointestinal anomalies can cause morbidity and mortality. In Japan, 10% of neonatal deaths were caused by congenital gastrointestinal anomaly surgeries performed. Abnormalities in newborns can be one type of abnormality or multiple congenital anomalies at the same time.<sup>9</sup> Osifo et al<sup>10</sup> reported that 30% of 78 neonatal deaths were due to congenital gastrointestinal anomalies, whereas our study found 27.6% of deaths. In the cases of diaphragmatic hernia, omphalocele, gastroschisis, and esophageal atresia, the highest mortality rates were from 2019 to October 2021 at Prof. Hospital. Dr. R. D. Kandou Manado. Cases of congenital anomalies affect 1 in 33 newborns, resulting in about 3.2 million people with disabilities each year. Despite the advances made in both the diagnosis and treatment, mortality of congenital anomalies remains most significant one of the most important causes of perinatal deaths. Several factors also influence the high mortality rate in neonates with gastrointestinal anomalies in developing countries, such as prematurity, comorbidities with other congenital anomalies, surgical complications, and inadequate Neonatal Intensive Care Unit (NICU) facilities.<sup>11</sup>

Sex differences in several specific congenital malformations have been documented as far back as the 1940s. Limited data is available concerning the sex distribution of

various congenital anomaly subtypes.<sup>2</sup> The study of Elghanni et al<sup>5</sup> reported on gender difference in congenital anomalies. The prevalence rate of congenital malformations was 1.02 %. There were 11 newborns that had sexual ambiguity, 395 were male and 276 were female. That study showed also that male newborns (57.9 %) had more congenital malformations than females (40.5%). The incidence of congenital malformations in our study was higher in males than in females. This is consistent with most studies worldwide. Albeit, some rare studies have shown a difference result with a higher prevalence of congenital malformations among females than males. Tennant et al<sup>12</sup> stated that male fetuses were significantly more prevalent in pregnancies affected by a congenital anomaly than female fetuses, but there was significant heterogeneity between subtypes ( $p < 0.001$ ). Forty-four of 110 (40%) unique subtypes were at least 40% more prevalent in males than females, with affected subtypes occurring across all major anomaly groups. Thirteen of 110 (12%) unique subtypes were at least 40% more prevalent in females than males, but the female-biased RR of a neural tube defect was less pronounced than previously reported.<sup>5</sup>

Until now, there is no definite data regarding the incidence of gastroschisis in Indonesia. However, the incidence of gastroschisis in developing countries is higher than in developed countries.<sup>1</sup> Age of pregnant women under 20 years is a significant risk factor for gastroschisis, especially in developing countries.<sup>13</sup> Mortality is also high in cases of low birth weight <2,500 grams and premature infants <34 weeks. Other mortality predictor factors that also play a role include pregnant women with diabetes mellitus since before pregnancy, and maternal obesity.<sup>8</sup>

Birth weight is a technically simple parameter to monitor prenatal health in a population. Establishing the prevalence of low birth weight (LBW) is particularly important, since perinatal morbidity and mortality are more frequent in LBW than in normal infants, and has become the second cause of death in this period after premature birth. In another study, Mekonen et al<sup>14</sup> reported that

of a total 1516 of deliveries, more female (54%) than male neonates were born. Birth weights were 700-1,000 grams between 26 and 36 weeks of pregnancy and then increased linearly to 3,500-4,000 grams at 40 weeks. Thirty-five and 54% of neonates were very low and low birth weight, respectively, without sex difference. Very low birth weight prevalence was not affected by parity. Male and female neonates from parity-2 and parity-2-4 mothers, respectively, were least frequently underweight. Sixty percent of newborns of parity -3 mothers weighed less than 2,500 grams without sex difference. The percentage of male neonates dropped from ~50% in parity-1-3 mothers to ~20% in parity-6 mothers. Diagnosed congenital malformations (~2%) were 2-fold more frequent in boys than girls. The commonest malformations were in the central nervous system (CNS; ~1.5% of newborns). Parity, low birth weight, gestational age less than 35 weeks, male sex, and lack of antenatal care were the most significant risk factors for congenital anomalies.<sup>15</sup>

In our study, duodenal atresia cases had the longest length of stay for  $\geq 14$  days. In the UK, 1 in 10 babies will require specialist neonatal care. Although the most preterm and smallest babies have the highest risk of mortality, if they survive their length of stay (LOS) in the neonatal unit will be very long. As neonatal survival has improved over recent years, particularly for very preterm babies, the number of babies requiring long-term neonatal care has increased. Genetic factors and disease factors during pregnancy are very influential in the occurrence of duodenum atresia. Other influencing factors include low birth weight, the presence of prematurity, or premature birth. The duration or length of treatment in the NICU takes a long time. It all depends on whether the general condition of the baby, namely the respiratory system, cardiovascular system, hematological system, gastrointestinal tract, nerves and other metabolisms are in a stable state and resolved or not. If all of these conditions can be resolved and the baby does not need supportive support, invasive oxygen therapy and then parenteral nutrition, the

baby can be cared for outside the NICU.<sup>11</sup>

## CONCLUSION

In this study, data were given in the form of risk factors related to the results of surgery for congenital anomalies, namely the case of recovery, male sex, aterm, birth weight  $\geq 2500$  grams, and length of stay for  $< 14$  days for the most in all cases at Prof. Dr. R. D. Kandou General Hospital Manado.

Hopefully, in the future, the service quality will improve in proportionate with increased number of surgical cases treated, and further studies with more specific numbers for each cases or risk factor will be produced.

## Conflict of Interest

The authors affirm no conflict of interest in this study.

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