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Relationship of Risk Factors for Pregnant Women with Congenital Disorders of Lobster Claw Syndrome, Ulutaue Hamlet, Mario Village, Mare District, Bone Regency, South Sulawesi

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ABSTRACT

The health condition of pregnant women is one of the risks that can increase the occurrence of congenital abnormalities; the prevalence of babies with congenital disorders in Indonesia is 59.3% per 1,000 live births. When compared to countries in Southeast Asia, Indonesia is still a country with a high prevalence of babies with congenital abnormalities. 50% of congenital abnormalities have no known cause; congenital abnormalities can be identified before birth, at birth, or later when the baby is born. Congenital abnormalities can affect organ shape, organ function, or both. Congenital abnormalities in infants vary from mild to severe and, in general, abnormalities occur in the first trimester phase, as happened in Ulutaue Village, there were some born with lobster claw syndrome, which they call stemmed fingers. This is a quite rare incident that occurs overseas only 1 in 90,000 live births, but it is different in Ulutaue Hamlet ranging from the elderly to children under five years old having fingers split in half, resemble crab claws. Most of the residents of Ulutaue Hamlet have suffered from this case. Reports in the last few years from the local health center found pregnant women giving birth with the same case. The purpose of this study is to determine the risk factors of pregnant women that cause the phenomenon of Lobster claw syndrome at Ulutaue Hamlet, Mare District, Bone Regency, South Sulawesi. This study uses a type of quantitative research with an analytical cross sectional design approach that aims at analyzing the relationship between risk factors of pregnant women and the occurrence of Lobster claw syndrome phenomenon.

Keywords: Pregnant Women, Lobster Claw Syndrome

INTRODUCTION

Congenital abnormalities can be identified before birth, at birth, or later after the baby is born. Congenital abnormalities can affect organ shape, organ function, or both. Congenital abnormalities in infants vary from mild to severe. The health and survival of babies with congenital disorders depend on the part of the body that has abnormalities. (Data and Information Center of the Indonesian Ministry of Health, 2018).

It is estimated 6% of infants worldwide are born with congenital abnormalities, resulting in hundreds of thousands of associated deaths. However, the actual number of cases may be much higher because statistics often do not take pregnancy terminations and stillbirths into account. WHO data states that from 2.68 million infant deaths, 11.3% are caused by congenital abnormalities (WHO 2016).

In Indonesia, the results of the Basic Healts Research 2017 explained that congenital abnormalities are one of the causes at infant mortality. In infants aged 0-6 days, infant mortality caused by congenital abnormalities was 1.4%, while at the age of 7-28 days, the percentage increased to

18.1% (Data and Information Center of the Indonesian Ministry of Health 2018). According to the global report on Birth released by the March of Dimes Birth Defect Foundation in 2006, the prevalence of babies with congenital abnormalities in Indonesia is 59.3 per 1,000 live births. When compared to countries in Southeast Asia, Indonesia is still a country with a high prevalence of infants with congenital abnormalities (March of Dimes 2006). Congenital disorders are an important health problem because they are one of the main causes of infant mortality in the world (Hoyert, D.L. and Xu 2012).

In South Sulawesi the number of neonatal deaths showed 799 cases with the neonatal mortality rate reach 5.30 per 1000 live births. In 2019 the number of neonatal deaths was 714 cases lower than the previous year with a neonatal mortality rate of 4.68 per 1000 live births (Public Health Office of South Sulawesi 2019).

Based on data from the Bone District Health Office, the number of neonatal deaths in 2019 was 63% (Bone District Health Office 2019). One of the rare congenital disorders in the world is "Lobster Claw Syndrome".

Definition of Lobster claw syndrome is a congenital absence of a central limb ray. The term lobster claw syndrome / ectrodactyly comes from the Greek words-ektroma (abortion) and daktylos (finger). Lobster claw syndrome (Ectrodactyly) was first documented in 1770 among the Guiana Indians. Von Walter described crab claw deformity in 1892, and Cruveilhier first used the term "lobster claw" in 1842. Ectrodactyly or Lobster Claws Syndrome can be caused by mutations in chromosome 7 and genetics. Lobster claw syndrome is also known as split hand and foot malformation (SHFM). This congenital abnormality is a rare disorder where only 1 in 90,000 live births are recorded abroad who experience the syndrome. However, this is different from one area in the village of Ulutaue, Bone, South Sulawesi, where the villagers have a disorder that is not common to humans (physically). The average human being is generally born with two hands, and has five fingers on each hand and foot. In Ulutaue Hamlet, the residents are born with their toes and hands split in half so that at first glance they look like crab claws which in medical language is called Lobster Claw Syndrome. The physical disorder has been hereditary. They have even given up because they believe that what they have experienced has been inherited by their ancestors.

LITERATURE REVIEW

1.General Overview of Pregnancy

Pregnancy is a very meaningful experience for women, families and societies, the behavior of the mother during her pregnancy will affect her pregnancy (Anita Widiastuti, et al 2021). A woman's health status before pregnancy will affect pregnancy; maternal health during pregnancy will affect her pregnancy and affect the growth and development of the zygote, embryo, and fetus including abnormal formation. The woman who is pregnant has previously suffered from a disease such as hepatitis, bladder infection, kidney disease, tuberculosis and others, so the midwife needs to reassess the woman's condition to find out if she is still facing problems related to the disease. This is very important because some diseases carried by the mother can have an impact on the baby they are carrying, such as syphilis or German measles which can cause birth defects (Anggita Sari 2015).

In general, the nutritional needs of pregnant women are the need for folic acid where folic acid is part of the vitamin B complex that can be isolated from green leaves such as spinach, fresh fruit, skin, liver, kidneys and mushrooms. Folic acid is also known as folacin / liver lactobacillus causal factor / factor U and factor R or vitamin B11. The need for folic acid up to 50-100 mg / day in normal women 300-400 mg / day in pregnant women while pregnant with twins is even greater. Folic acid deficiency causes placental disorders, habitualist abortion, placental abruption, and congenital

abnormalities in the fetus. (Anggita Sari 2015)

2.General Overview of Congenital Abnormalities

According to the International Classification of Disease Tenth Revision (ICD10) birth defects are congenital abnormalities that include congenital malformations, deformations and chromosomal abnormalities (WHO 2012). Congenital disorders are an important health problem because they are one of the causes of infant mortality in the world (Hoyert, D.L. and Xu 2012). Congenital disorders are a health burden for countries with low to middle income even some countries do not have adequate service facilities to deal with these occurrence so that they can have long-term effects. WHO: of all infants with congenital abnormalities, more than 70% die in the first month (Rizk Francine, Salameh Pascale 2014). The exact etiology of this congenital abnormality is not known. About 40%-60% cases have no known cause, but it is estimated that gene mutations are the most influential factor in the occurrence of congenital abnormalities (Căpățînă and Cozaru 2015).

Among several etiologies of genetic disorders, chromosomal abnormalities reach 6%, single gene disorders 25%, and multifactorial 20-30%. Same-sex marriage is considered as one of the important factors that contribute to the incidence of congenital abnormalities (Rizk Francine, Salameh Pascale 2014). There are several risk factors associated with congenital abnormalities, namely maternal age, multiparity, history of abortion, congenital abnormalities in previous pregnancies, gestational diabetes, exposure to cigarette smoke, alcohol consumption, drug consumption, not taking folic acid, family history of abnormalities congenital, there is a blood relationship between father and mother, and low socioeconomic status (Febriano Matthew 2021). Genetic disease in the perinatal period is a disorder that can manifest from conception to birth with symptoms of infertility, abortion, fetal death, and disease or disability in neonates, infants and children (Wiknjosastro 2010). One of the congenital disorders caused by genetic disorders is EEC syndrome which is a rare autosomal dominant genetic disorder characterized by varying degrees of ectrodactyly and syndactyly (hands and feet) (Rudiger RA 1970). Most cases of EEC syndrome are caused by heterozygous mutations in the p63 gene (Brunner, Hamel, and Van Bokhoven 2002).

MATERIALS & METHODS

This research design used in this study is observational analytic cross-sectional study design, which aimed at analyzing the relationship between risk factors of pregnant women and the occurrence of lobster claw syndrome. The data used in this study is primary data by visiting the respondent's home.

A. Operational Definition and Measurement

1.Syndrom lobster claw

Operational Definition

Lobster claw syndrome by strict definition is the congenital absence of a central ray of the limbs. The term lobster claw syndrome / ectrodactyly comes from the Greek words-ektroma (abortion) and daktylos (finger)

Objective Criteria:

Syndrome lobster claw: If pregnant women with signs of not having one of the fingers or legs on the feet

Don't have lobster claw syndrome: If pregnant women do not have signs of lobster claw syndrome

Inbreeding/Marriage between Relatives

Operational Definition

Inbreeding is a marriage between a woman and a man who are still closely related by blood or still in relatives.

Objective Criteria:

Inbreeding: a marriage between a woman and a man who are still closely related by blood or still in relatives.

Not Inbreeding: A Marriage between a woman and a man that is not closely related by blood

1.Anemia

Operational Definition

Anemia is defined as a low concentration of hemoglobin (Hb) in the blood (WHO 2015)

Objective Criteria:

Anemia: defined as a low concentration of hemoglobin (Hb) in the blood (WHO 2015)

Not Anemia: normal concentration of hemoglobin (Hb) in the blood

2.Age of Marriage

Operational Definition

The age of marriage is regulated in Law Number 16, 2019. Marriage is only permitted if the man and woman are 19 years old

Objective Criteria:

High- risk marriage age :<19 >30Years old

Low – risk marriage age: 19-30Years old

3.Nutritional Status

Operational Definition: The nutritional status of pregnant women is a state of balance in the body of pregnant women as a result of the intake of food consumption and the use of nutrients used by the body. The nutritional status of pregnant women can be known by measuring the upper arm circumference (LILA).

Objective Criteria:

Pregnant Women with Chronic Energy Deficiency : Pregnant women who have LILA size < 23.5 cm

Pregnant Women with Not Chronic Energy Deficiency: Pregnant women who have LILA size >23,5cm

Statistical Analysis : Data analysis used univariate analysis and bivariate analysis.

RESULT

1.Relationship of inbreeding factors with the occurence of lobster claw syndrome

Based on the results of the study, the results of the chi square test on inbreeding with the occurrence of lobster claw syndrome p value = 0.017, which means that there is a relationship between inbreeding and the incidence of lobster claw syndrome. Most of the scientific literature on cousin marriage is concentrated on quite specific aspects of the effect of inbreeding on fertility and health (Bittles and Black, 2010: 197) in (Yusdiawati 2018) (Hamamy 2012) conducted a consequence study group of international experts and counselors conducted at the Geneva International Consanguinity Workshop from 3 May 2010 to 7 May 2010, to discuss the suspected risks and benefits of close family marriage. They found there are health risks for couples in cousin marriage, especially in Middle Eastern states.

Relationship between Married Age and Lobster Claw Syndrome

Based on the results of the study, the results of the chi square test on the age of marriage with the occurrence of lobster claw syndrome p value = 0.92, which means that there is no relationship between the age of marriage and the occurrence of lobster claw syndrome. This is in line with research (Ellyati, Kusharisupeni, and Sabri 2019) with the title Factors related to congenital abnormalities with a chi-square statistical test value obtained p value = 0.309 meaning that there is no relationship between maternal age and the incidence of congenital abnormalities.

2.The relationship between HB levels of pregnant women and the occurence of lobster claw syndrome

Based on the results of the study, the results of the chi square test on hemoglobin levels with the incidence of lobster claw syndrome p value = 0.014, which means that there is a relationship between hemoglobin levels and the occurence of lobster claw syndrome.

3.The relationship between the nutritional status of pregnant women and the incidence of lobster claw syndrome

Based on the results of the study, the results of the chi square test on the nutritional status of pregnant women (upper arm circumference) with the incidence of lobster claw syndrome p value = 0.017, which means that there is a relationship between nutritional status and the incidence of lobster claw syndrome. The nutritional status of the mother before pregnancy and during pregnancy can affect the growth of the fetus being conceived. Pregnant women

who suffer from CED and anemia have a greater risk of illness, especially in the third trimester of pregnancy compared to normal pregnant women. This is in line with research (Lubis 2003) that there is an impact of malnutrition on pregnant women on their fetuses, which can affect the process of fetal growth and can cause miscarriage, abortion, stillbirth, neonatal death, and congenital defects.

CONCLUSION

Based on the results of research conducted in Ulutaue Hamlet, Mario Village, Mare District, Bone Regency, with analysis using the Chi square test and the level of significance (α) 0.05, it can be concluded that the occurrence of lobster claw syndrome is related to the inbreeding variable as evidenced by the statistical test results p value <0.017, and is also associated with hemoglobin levels of pregnant women with test results chi square p value 0.014. There is also a relationship between the occurrence of lobster claw syndrome and the nutritional status of pregnant women with a chi square value of 0.017 and there is no significant relationship to the variable age of marriage with the results of the chi-square test p value > 0.92.

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