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Clinical and laboratory features of childhood systemic lupus erythematosus and its course in Department of Child Health, Cipto Mangunkusumo Hospital, Jakarta

Zakiudin Munasir, Tuty Mariana, Rulina Suradi

Department of Child Health, Faculty of Medicine, University of Indonesia, Cipto Mangunkusumo Hospital, Jakarta

ABSTRACT A descriptive, retrospective study has been performed among 33 children with SLE during the period of January 1986 to December 1999 in Department of Child Health, Cipto Mangunkusumo Hospital. The results of the present study show that SLE in childhood was more commonly found in girls than in boys, with a ratio of 4.5: 1. The mean age of onset was 10.5 years and the mean age of diagnosis was 10.9 years, most frequently observed in the age of > 10 years and rarely found in the age of < 5 years. The time interval between the time of onset and time of diagnosis ranged from 1 to 24 months and the most frequent interval was 1-3 months. The most common initial symptoms were prolonged fever, rash on the skin and face, and athralgia. In its natural history of the disease, kidneys, skin/mucous membrane and joints are the most frequent organs involved. Most of the SLE patients develop anemia. Positive anti ds-DNA, ANA and decreased levels of C3 and C4 respectively in 28 (28/31), in 29 (29/30), in 25 (25/33), and in 19 (19/27) cases. LE cells were encountered only in four (4/17) cases. Out of 11 cases upon which renal biopsies were done, the most common histological features were mesangeal glomerulonephritis (class II) and diffuse proliferative glomerulonephritis (class IV). The mean time interval between the onset and renal complication manifestations was 6.96 months, cardiac complication was 16.77 months, central nerve system was 22.71 months and lungs were 25.0 months. Duration of illness of patients with SLE ranged from 2 to 175 with the mean of 31.3 months. The causes of death were mostly due to gastrointestinal bleeding and renal failure. **[Paediatr Indones 2001; 41:214-224]**

Keywords: systemic lupous erythematosus, clinical course, children, antinuclear antibody

SLE IS A MULTI SYSTEM DISEASE CHARACTERIZED BY extensive inflammation of blood vessels and connective tissues. [1,2] The condition is rarely found in childhood, generally seen in adolescence and girls are more frequently affected than boys.[1-4]

The etiology of SLE has not been known exactly, and the pathogenesis is thought to begin with incapability of the immune system to recognize the structure of self-antigen so that autoimmune mechanism occurs. [1.5]

The clinical manifestations of SLE vary greatly, the initial symptom does not necessarily show multi organ involvement. The initial symptoms often have manifest long time before the diagnosis of SLE. Generally the clinical course of SLE, is chronic in nature, hard to suspect, still difficult to be treated, frequently progressive and ends with death.[1, 2]

Up to now, in Indonesia reports of' researches concerning SLE disease in childhood are rare. The

Correspondence: Zakiudin Munasir, M.D., Department of Child Health, Medical School, University of Indonesia, Jalan Salemba 6, jakarta 10430. Tel 62-21-3907742, Fax 62-21-3907743

current study aimed to identify the clinical, laboratory features of the condition and its clinical course in children in the Department of Child Health, Cipto Mangunkusumo Hospital.

Methods

This was a descriptive, retrospective study performed on 33 children with SLE extending from January 1986 to December 1999 in the Department of Child Health, Cipto Mangunkusumo Hospital. The data was taken from the medical records. Diagnosis of SLE was made according to ARA criteria revised in 1982,[1] except for proteinuria which was determined >1+ (> 30 mg) using dipstick tests more than once.[6-8] Age of onset was age when developed symptoms based on interview and/or the referring information.[9, 10] Age of diagnosis was age of patient fulfilling 4 or more out of 11 criteria of ARA (revised in 1982).[9] Renal manifestation included abnormalities in urinalysis (proteinuria, hematuria), renal microscopic abnormalities on specimen biopsy.[11] Hematuria when red blood cells of more than 2/hemoglobin/granular/tubular were found in urine.[8] Cardiac,[12] lungs[13-15] and central nerve system (CNS)[1] manifestations included clinical symptoms/signs combined with supporting examination results. Histopathologically, lupus nephritis was in accordance with WHO classification.[1,16] The criteria of anemia was Hb<10g%, leukopenia was <4000/ul, and thrombocytopenia when the thrombocyte count was <100.000/ul.[4,9] The C3 and C4 levels was measured with ELISA method. Normal value of C_3 was 55-120 mg/dl, and C_4 was 20-50 mg/dl. Immunological disorder was defined when titer of ANA, anti ds-DNA and LE cells was positive. ANA study was conducted with immunofluorescence. The duration of illness was the length of patients suffering from SLE since the patients developed the onset until the end of the study or if she/he died.

Results

Extending from January 1986 to December 1999, 37 children with SLE were hospitalized. (See Figure 1). However, only 33 medical records could be found.

N = 37

Subject Characteristics

Out of 33 cases under investigation, 27 were girls and six were boys, the ratio of girls to boys was 4.5: 1. The mean age of onset was 10.5 years; the mean age at diagnosis was 10.9 years. The age of onset and age at diagnosis was most common in the age of ≥ 10 years. See Table 1.

TABLE 1. CHARACTERISTICS OF STUDIED SUBJECTS

Variable	N=33	Mean (SD)	Range
Sex			
Female	27		
Male	6		
Age of onset (years)			
< 5	1	10,5 (± 2,2)	4,6 – 15,4
5 - < 10	12		
≥ 10	20		
Age of diagnosis (years)			
< 5	1	10,9 (± 2,2)	4,8 – 15,9
5 - < 10	9		
≥10	23		

Interval time between the onset and diagnosis of SLE

Table 2 shows the interval time between the developments of onset up to the time of diagnosis of SLE. It ranges from 1 to 24 months, and the most common interval time was 1-3 months.

Clinical Features

At the time of onset, the most frequent observed symptoms were prolonged fever (fever lasting for more

TABLE 2. INTERVAL TIME BETWEEN THE ONSET AND DIAGNOSIS OF SLE

Interval time between onset and diagnosis of SLE (months)	N=33	
1 - 3	20	20/33
> 3 - 6	6	6/33
> 6 - 9	3	3/33
> 9 - 12	1	1/33
> 12 - 15	0	
> 15 - 18	2	2/33
> 18 - 21	0	
> 21 - 24	1	1/33

than 2 weeks), followed by rash on the skin/face, and athralgia. Six cases with symptom of pallor accompanied by positive evidence of Coombs' test in one case. Four cases with edema at onset have been with lupus nephritis. In its natural course, SLE disease frequently shows fever, rash on the skin/face and athralgia persistently. (See Table 3).

Table 4 indicates organ involvement during the clinical course of SLE. The most frequently involved organ is the kidney.

Laboratory Findings

Table 5 shows that anemia occurred in 28 cases, and of 16 cases upon whom Coombs' tests were conducted, five of them showed positive results. Leukopoenia was encountered in only nine cases. Thrombocytopenia was observed in five cases and all of them developed bleeding, three of the five died of hemorrhage. In this investigation the serological studies of ANA, anti ds-DNA, C3 and C4 also LE cells can be seen in Table 5.

Histopathological features of the kidney towards ANA, anti ds-DNA, C3 and C4, renal function, cause of death and duration of illness in 11 patients with SLE.

Out of 31 cases with renal manifestation, only 11 cases were done renal biopsy. The most prevalent histopathological features were proliferative glomerulonephritis (class IV) in three cases and mesangeal glomerulonephritis also in three cases (class II), whereas membranous glomerulonephritis (class III) was found in one case and normal glomerulus (class I) was found in one case. All cases (3 patients) with histopathological features of class IV (diffuse proliferative glomerulonephritis) died, two of them were known to die of terminal chronic renal failure, while the other one case the cause of death was unknown. All cases (3 patients) with histopathological features of class II (mesangeal glomerulonephritis) also died, but the cause of death was undetermined. In one case, the renal histopathologiocal feature was not representative, but at the end of the study she was known to have with terminal chronic renal failure and was undergoing hemodialysis.

Table 6 depicts positive anti ds-DNA in the entire cases but in two cases, the results were not known because of unavailable data. Negative ANA was observed in one case. Serum concentration of C_3 declined in nearly all cases, but in one case with histopathological features of class IA showed normal serum concentration of C_3.

Interval time between the onset until the development of renal, cardiac, pulmonary and central nerve system complications.

The presence of renal, cardiac, central nerve system and pulmonary complications can be determined by its clinical manifestations and can be proven with supporting examination results. Table 7 indicates that the interval time between the onset and the development of complications. Duration of illness

The range disease duration of SLE patients in the present study was from 2 to 175 months with the mean of 31.3 months, the most common range was between 2 to 12 months in 14 cases, and 12 of them died. All male cases died with duration of illness less than 36 months.

TABLE 3. SIGNS/ SYMPTOMS OF THE ONSET AND DURING THE CLINICAL COURSE OF SLE.

Signs/ symptoms	At onset N=33	During the course of the disease N=33
Fever	33	33
Rash on skin/ face	28	28
Athralgia	23	24
Fatigue	21	27
Anorexia	20	27
Lost of weight	15	4
Alopecia	10	17
Stomatitis	6	19
Nasal ulceration	0	1
Pallor	6	25
Edema	4	9
Lymphadenopathy	3	9
Hepatomegaly	0	14
Splenomegaly	0	3
Arthritis	2	7
Pleural effusion	0	5
Pneumonitis	0	3
Seizures	0	15
Encephalopathy	0	6
Psychosis	0	4
Intracranial bleeding	0	1
Pericarditis	1	10
Myocarditis	0	3
Endocarditis	0	1
Hypertension	0	11
Gastrointestinal bleeding	1	8
Pancreatitis	0	6
Diarrhea	0	3
Conjungtivitis	0	2

TABLE 4. ORGAN INVOLVEMENT DURING THE COURSE OF THE DISEASE

Organ involvement	N=33
Renal	31
Skin and mucous membrane	30
Joint, bone and muscle	24
Cardiovascular	19
CNS	17
Liver, spleen, lymph nodes	14
Gastrointestinal tract	14
Lungs	6
Eyes	2

TABLE 5. LABORATORY FINDINGS OF PATIENTS WITH SLE

Laboratory findings	N=33	
Anemia	28	28/33
Coomb's test positive	5	5/16
Coomb's test negative	11	11/16
Leukopenia	9	9/33
Thrombocytopenia	5	5/33
Erythrocyte sedimentation rate elevated	33	33/33
Urinalysis abnormal	31	31/33
Proteinuria	31	31/33
Hematuria	12	12/33
ANA positive	28	28/33
Anti ds-DNA positive	29	29/33
LE cell positive	4	4/33
Complement C3		
Decrease	25	25/33
Increase	6	6/33
Complement C4		
Decrease	19	19/27
Increase	6	6/27

Cause of death

Out of 33 cases under study, 24 cases died 12 cases died in Cipto Mangunkusumo Hospital and the direct cause of death could be identified. Of these 12 cases, the most prevalent cause of death was gastrointestinal bleeding and renal failure.

Discussion

Data obtained in the study were based on secondary data from medical records in the Department of Child Health, Cipto Mangunkusumo Hospital, during the period of 15 years (from January 1986 to December 1999). Using a descriptive, retrospective design, in the present study some limitations of assessment, such as incomplete data, uniformed treatment protocol and incomplete information obtained from this study made analysis difficult.

From January 1986 to December 1999 from the medical records in the Department of Child Health, Cipto Mangunkusumo Hospital, the data of 37 cases were recorded. Figure 1 shows data of new patients with SLE in 1986 – 1999 in the Department of Child Health, Cipto Mangunkusumo Hospital. From January 1986 to December 1989 the number of new patients were only 1 to 2 cases annually, maybe because the diagnosis of the disease is not recognized widely. However, after the year of 1990, as the diagnostic technology became advanced and increased awareness among medical doctors toward the diagnosis of the disease, the number of patient increased and the largest number was found in 1995 and 1996. In 1998, the number of cases seemed to decline that might have been because of economic situation in Indonesia being in crisis, which made the ability to visit a hospital decrease among the low-income society.

After the data collection from medical records, only 33 of cases could be obtained. Table 1 shows that the number of girls was 27 and boys were six, with girls and boy's ratio was 4.5:1. Similar with the reports by Norris et al, [4] the ratio was 5.7:1 Fish et al, [17] and Font et al, [9] reported the girl and boys ratios were 11:1 and 10:1, respectively.

In this study the mean age of onset was 10.5 years with range of 4.6-15.4 years, the most prevalent age was $\geq$ 10 years, and rarely encountered in the age of <5 years. These results were in accordance with that reported by Norris et al [4] and Caeiro et al.[18] In this study the mean age when diagnosis made was 10.9 years with a range of 4.8 to 15.9 years, and the most frequent age found was $\geq$ 10 years. The study by Fish et al [17] showed that the mean age at diagnosis of SLE was 12.9 years with range of 2.2 to 18 years and Norris et al [4] reported the mean age of 11.7 years with 2-15 year range.

Table 2 shows that the initial symptoms in SLE patients have manifested long before the diagnosis was established. In one case, the initial symptom occurred 24 months before, and two cases showed initial symptoms for 16 - 18 months before the diagnosis of SLE was made. Most cases had developed the initial symptoms from 1 to 3 months before the diagnosis. Other researchers reported the same results. [19]

In the present investigation, the most common initial symptoms were fever, rash on the skin/face and athralgia and they persisted during the clinical course. Followed by cases with symptoms of fatigue, anorexia, alopecia, stomatitis, pallor and pericarditis increased. Another study also reported the most frequent encountered symptoms of fever, rash on the skin and joint pain.[17,18,20,21] Therefore, if there were symptoms of fever, rash on the skin/face and joint pain the possibility of SLE disease should be considered and needs further monitoring.

Table 4 shows that the organs most frequent involved were the kidneys (31 cases). In the literature, it was reported that renal involvement occurred more often in SLE with childhood onset than in adult onset. In the present study renal involvement had been seen in initial course of 4 cases Font et al, [9] reported such involvement as many as 50% of cases and renal manifestations had been observed in initial natural course in 20% of cases. Phadke et al [22] reported two cases with manifestation of acute renal failure as the initial manifestation of SLE. Leng [23] in Singapore made report of renal involvement in childhood SLE in 74% of cases, likewise Fish et al [17] in Sweden was 77.5%, Baron et al [24] in Canada was 82%. Because of the very high frequency of nephritis lupus in childhood SLE, the SLE patients with absent or later urinalysis abnormalities were called silent lupus nephritis. Therefore, periodical renal evaluation should be conducted in all SLE patients.

Previous studies reported that activation of renal abnormalities was associated with increased anti ds-DNA and decreased levels of serum complement.[25, 26] In this study, in Table 6 shows that anti ds-DNA increased and serum concentration of complement declined, indicating that lupus nephritis was still active.

When associated with renal histopahtological features (Table 6), three cases with renal histopathological features of class IV (diffuse proliferative glomerulonephritis) died. Two of them died with terminal renal failure. The other, but on the last visit has been known as experiencing chronic renal failure. Some studies showed that in SLE with renal abnormalities, the predominant cause of death in-patients with clinical presentation of glomerulonephritis or histopathological features of diffuse proliferative glomerulonephritis, the main cause of death was terminal renal failure.[20.27]

TABLE 6. RENAL HISTOPATHOLOGY FINDINGS WITH ANA, ANTI DS-DNA, C_3-C_4 COMPLEMENTS, RENAL FUNCTIONS AND THE CAUSE OF DEATH OF 11 SLE PATIENTS.

No. cases	Histology (WHO)	Proteinuria	Ureum/creatinin (mg/dl)	Creatinine clearance (ml/mean/1,73 M^2)	C_3	C_4	ANA	Anti ds-DNA	Survive /died	Cause of death	Duration of illness (years)
2	Class V	3+	25/0.56	n.d	↓	↓	+	n.d	Survive	-	14.42
3	Class IA	3+	20/1.60	n.d	↓	n.d	+	+	Survive	-	0.33
5	Class IV	4+	661/5.93	6.23	↓	n.d	n.d	+	Died	Terminal renal failure	6.16
9	Class IV	3+	60/6.23	n.d	↓	↑	+	+	Died	?	3.00
10	Class IV	3+	176/2.50	1.96	↓	↓	+	n.d	Died	Terminal renal failure	0.33
14	Class III	3+	99/0.8	n.d	↓	↓	+	+	survive	-	4.16
19	Class IA	2+	21/0.6	n.d	N	n.d	+	+	Survive	-	0.92
22	Class II	3+	147/4.29	25.15	↓	↓	n.d	+	Died	?	2.50
23	Class II	3+	18/0.82	27.8	↓	↓	-	+	Died	?	1.58
28	Class II	2+	34/0.90	n.d	↓	↓	+	+	Died	?	3.66
8	No representative	3+	262/13.44	13.44	↓	↓	+	+	Survive	-	12.08

The other organs frequently involved were the skin and mucous membranes. One of the SLE characteristics is photosensitivity due to the skin exposure to ultraviolet rays, so SSA antigen expression in keratinocyte surface increases. The binding of autoantibody to this keratinocyte SSA antigen will increase complement-making inflammation that gives abnormalities in the skin.[1]

Joint involvement often occurs in SLE. In the present study, joint disorders (athralgia and arthritis) at the onset of the disease were observed in 23 cases and in its clinical course took place in 24 cases. Norris et al [4] reported joint disorders (athralgia and arthritis) at onset took place in 47.52% of cases and in its clinical course were observed in 66.40% of cases. Meanwhile Font et al [9] reported at onset joint disturbances occurred in 65% of cases and during the clinical course were observed in 88% of cases. Unlike rheumatoid arthritis, joint abnormalities occurring in SLE, generally, do not result in bone erosion (joints deformities occurred were reversible). [28]

Table 4 shows liver, spleen and lymphatic involvement in 14 cases. Hepatomegaly in children with SLE usually is rare, jaundice is rarely related to the presence of liver abnormalities and ordinarily icterus occurred in association with hemolytic process. Splenomegaly occurred in relation to active SLE and usually in moderate grade. Lymphadenopathy also was seen in the acute phase of the disease and frequently found in children than in adult and markedly decrease was observed after treatment.[1]

Another organ involved in SLE, though rare, is the central nerve system. In this study, manifestations of the CNS initially had not been observed, but in its clinical course it was seen in 17 cases as seizures, encephalopathy, psychosis and intracranial bleeding. Unlike the study reported by Fish et al, [17] central nerve system manifestations had been observed both since the onset and during its clinical course (in the therapy period). Initially, it was thought that the chief cause of central nerve system abnormalities was vasculitis. However, in the present, another mechanism thought as the cause of CNS abnormalities is thrombosis.[29-31]

Pericarditis was the most common cardiovascular manifestation. Another researcher also reported the same results.[4, 18] In this study all patients developed hypertension accompanied with renal disorders. Hypertension is caused by the presence of lupus ne-

TABLE 7. THE MEAN TIME FROM THE ONSET UNTIL THE PRESENT COMPLICATIONS OF PATIENTS WITH SLE.

Complication	N	Duration until the complications occurs		
		Mean (months)	Standard deviation (months)	Range (months)
Renal	31	6,96	6,17	1-24
Cardiac	13	16,77	19,77	1-62
CNS	17	22,71	19,29	2-66
Lungs	6	25,00	26,20	1-62

TABLE 8. DURATION OF ILLNESS OF PATIENTS WITH SLE

Duration of illness (months)	Survive		Died		Total
	M	F	M	F	
2 - 12	0	2	3	9	14
>12 - 24	0	1	2	2	5
>24 - 36	0	1	1	2	4
>36 - 48	0	0	0	1	1
>48 - 60	0	1	0	1	2
>60 – 120	0	2	0	3	5
>120	0	2	0	0	2
Total	0	9	6	18	33

phritis and by corticosteroid treatment.[32]

In the current study gastrointestional involvement occurred in 14 cases, as gastrointestinal bleeding, pancreatitis and diarrhea. Gastrointetinal disturbances could be due to vasculitis that can cause gastrointestinal bleeding, pancreatitis, intestinal perforation and hemorrhagic ulceration. Tsukura et al [33] reported the presence of protein losing enteropathy and malabsorption syndrome with diarrhea in-patients with SLE, this was related to treatment with salycilic acid, corticosteroid, antimalarial and cytotoxic agents.

Pulmonary involvement in childhood SLE rarely occurs, the most common lung manifestations were pleural effusion mainly in active SLE.[1, 13] In this study pulmonary involvement took place in 6 cases, and the most common lung manifestations were pleural effusion and pneumonitis consecutively occurred in 4 cases and in 3 cases. Another study found the same results.[4, 9,13] The presence of lung abnormality in SLE is considered if the underlying infection is excluded.[1]

The eye abnormalities are rarely reported. In this study, ophthalmologic abnormalities were only observed in two cases as conjunctivitis. Conjunctivitis in SLE patients can be due to secondary infection both by virus and bacteria, can also be a clinical manifestations of conjunctival vasculitis.[34]

Table 5 shows that most patients with SLE develop anemia. Anemia in SLE can be caused by hemolytic process, because of the presence of autoantibody towards erythrocytes, which was proven by a positive Coombs test, which was found in five cases (out of 15 case upon which Coombs test was performed). But anemia can also be caused by non-immune processes such as blood loss including menorrhagia and gastrointestinal bleeding, iron deficiency, chronic clinical course, and bone marrow depression due to cytostatic drugs and due to chronic renal failure. Caerio et al [18] reported anemia in 52.3% of cases with positive Coombs test in 7 out of 11 cases undergoing Coombs test. Norris et al [4] reported anemia in 63.4% cases, hemolytic anemia in three cases with positive Coombs test. Font et al [9] reported hemolytic anemia in 15% cases. However, Fish et al [17] reported hemolytic anemia with positive Coombs test in 73% cases.

Leukopenia (leukocytes < 4.000 cells/ul) was an important marker in acute SLE. In this study, it was encountered in eight cases. The study results were almost the same as those reported by other researchers.[4, 17,18] The low number of leukocyte leads to the susceptibility to infection. Leukopenia is associated with disease activity and the presence of autoantibody towards lymphocyte.

Bleeding due to thrombocytopenia can occur in SLE. In the present study thrombocyte of <100.000/ul was found in 5 cases and result in bleeding in the five cases, 3 of them died of bleeding. Hemorrhage in SLE can also be due to the presence of clotting factors disorder, thrombosis or drugs. Thrombocytopenic purpura accompanied with hemolytic anemia (Evan's syndrome) as reported by Norris et all in 2 cases SLE[4] was not observed in this study. In the present study all SLE patients showed increased erythrocyte sedimentation rate (Table 5), this was in accordance with the presence of inflammatory process occurring in SLE patient. [35]

In the present study, the operational definition of protenuria was +1 or more (30 mg or more), as mentioned in the literature that patients with renal anatomic pathological features of class II and I (WHO criteria) shows mild clinical presentation namely normal urinalysis or minimal and normal renal function.[36] Table 5 shows abnormal results of urinalysis in 31 cases, as protenuria (31 cases) and hematuria (12 cases).

TABLE 9. THE CAUSE OF DEATH OF 12 PATIENTS WITH SLE AT CIPTO MANGUNKUSUMO HOSPITAL.

Cause of death	N=12
Gastrointestinal bleeding	4
Terminal renal failure	2
Renal failure	1
Septic shock	1
Hypovolemic shock due to DIC	1
Encephalopathy	1
Cardiac failure	1
Aspiration	1

This abnormal urinalysis indicates the presence of lupus nephritis.

In the study positive ANA and anti-ds- DNA were observed in 28 (28/31) cases and 29 (29/30) cases, respectively while levels of C3 and C4 decline in 25 (25/33) cases and 19 (19/27) cases, suggesting that the disease was still active. It was stated in the literature that positive ANA was found in almost all children with active SLE.[37] The ANA studies, therefore, should always be performed in cases with suspicion of SLE. The autoantibody ANA is a highly sensitive examination, but this method is not necessarily specific because ANA can also be encountered in other diseases such as Sjogren's syndrome, scleroderma and rheumatoid arthritis.[38] The more specific antibody for active SLE is anti ds-ANA. In SLE patients with active renal abnormalities anti ds-DNA incessantly increases. Hypocomplementemia was observed in increased anti ds-DNA at once.[38] Singseng et al [39] reported various manifestations in childhood SLE can result in hypocomplementemia. Hypocomplementemia is not seen only in active lupus nephritis but also in-patients with active skin and central nerve system manifestations.[39]

Table 7 shows interval time between the onset until the development of renal complication ranging from 1 to 24 months with the mean of 6.96 months. Four cases had been with lupus nephritis at the time of diagnosis made. According to the literature, lupus nephritis in childhood more frequently occurs in the first years of clinical course of lupus.[1, 22] The interval time between the onset until cardiac complication development ranged from 1 to 62 months with the mean interval was 16.77 months. One case on admission had pericarditis before the diagnosis of SLE was established. Interval time between onsets until the development of complication in CNS ranged from 2 to 66 months with the mean interval of 22.71 months. Cassidy et al [18] reported that CNS complications could occur at any time during the clinical course. In children this CNS complications can appear after the commencement of treatment.

The duration of illness in SLE patients is determined from the development of onset based on medical interview up to the end of study or at the time of death. In the present study, the mean duration of illness of patients with SLE was 31.3 months with range of duration of illness from 2 to 175 months. The most common duration of illness was between of 2 to 12 months in 14 cases, 12 of them died. Font et al [9] reported the mean duration of illness in childhood was 85 months with range time of 1 to 264 months. In the current study duration of illness in boys was not more than 36 months and all cases (6 patients) died, suggesting that SLE in boys was more severe. This was similar with the study by Celermajer et al.[7]

It is stated in the literature, the cause of death can be directly due to lupus disease or caused by side effects of treatment or due to immune deficiency resulted from lupus disease.[1] Renal involvement is the main cause of death in childhood SLE. CNS involvement is the second cause of death.[21] In this study out of 33 cases under investigation, 24 cases died, but the direct cause of death known was only in 12 cases, in the remaining the cause of death was unknown exactly because the patients died at home. The most frequent cause of death was due to gastrointestinal bleeding (4 cases), followed by renal abnormalities (3 cases). The cause of death related to CNS involvement was found in one case, due to heart failure in one case, DIC in one case and septic shock in one case. Gastrointestinal bleeding in SLE is a serious condition that may result from the lupus disease itself, or due to side effects of treatment.

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